COCHLEAR IMPLANTS IN ADULTS

Results from the Nijmegen-Sint Michielsgestel Cochlear Implant Program

Johannes B. Hinderink

COCHLEAR IMPLANTS IN ADULTS

Results from the Nijmegen-Sint Michielsgestel Cochlear Implant Program

een wetenschappelijke proeve op het gebied van de Medische Wetenschappen

Proefschrift

ter verkrijging van de graad van doctor
aan de Katholieke Universiteit Nijmegen,
volgens besluit van het College van Decanen in het
openbaar te verdedigen op maandag 26 maart 2001
des namiddags om 3.30 uur precies

door

Johannes Barabbas Hinderink

geboren op 10 april 1962
te Den Haag

Promotor:	Prof. Dr. P. van den Broek
Co-promotor:	Dr. L.H.M. Mens
Manuscriptcommissie:	Prof. Dr. P.F. de Vries-Robbé, voorzitter Prof. Dr. A.F. Deutman Prof. Dr. F.J.M. Gabreëls

Omslagillustratie uit *Theatrum Universale* door John Jonston, 1660. Kopergravure door Matthias Merian

Printed by Print Partners Ipskamp, Enschede

CIP-gegevens Koninklijke Bibliotheek, Den Haag

Hinderink, J.B.
Cochlear Implants in Adults: Results from the Nijmegen-Sint Michielsgestel Cochlear Implant Program
Proefschrift Katholieke Universiteit Nijmegen
- Met literatuuropgave - Met samenvatting in het Nederlands

ISBN 90-9014632-6

TABLE OF CONTENTS

CHAPTER ONE

COCHLEAR IMPLANTATION: AN INTRODUCTION

INTRODUCTION

Until a few decades ago, total deafness was incurable. Those affected depended very much on communication by lip reading and gestures, perhaps supplemented by instruments to make it possible for sounds to be felt.[1] Auditive contact with the surrounding environment was not possible.

Cochlear implantation (CI) has fundamentally altered this situation. CI makes it possible to perceive sounds through direct electric stimulation of the auditory nerve through electrodes placed in or nearby the cochlea. Cochlear implantation began around 1960 and has been applied clinically on a large scale since the 1980s. Many studies have since been conducted to take stock of results gained with CI in adults, mostly in the form of research into the effect on speech perception.[2,3,4,5]

Initially, only post-lingual deaf adults were implanted. Rapid technological development and favorable results have, however, increasingly allowed for cochlear implantation in deaf children in the past ten years. In the meantime research has shown that CI has had not such a favourable outcome for pre-lingual deaf adults, probably due to physiological factors such as insufficient maturity (involution of the auditory system) although social factors also play a role. Pre-lingual deaf adults are usually members of deaf communities with alternative full-scale communication methods, such as sign language, available. Nevertheless, CI may be indicated for a small group of pre-lingual deaf adults including those with, for instance, an additional visual handicap such as in the Usher syndrome.[6]

As an introduction to the present study, a short description of the normal anatomy and physiology of the ear will be given together with a description of some pathological features of deafness. This will be followed by a description of the working principle of a cochlear implant and a review of various implant systems. The introduction will conclude with a short overview of the situation in the Netherlands regarding selection procedure, implantation, rehabilitation, and evaluation of results. Before this, a review of the history of CI is in order.

HISTORY OF CI

Graeme Clark, a pioneer of CI, provides an outstanding summary of its early history in his dissertation dating from 1969.[7] The following is largely drawn from his presentation.

The initial description of the electric stimulation of the auditory system was provided in 1800 by Count A. Volta, the inventor of the electrolytic cell (1790). Upon placing metal electrodes in his ears and connecting a current of about 50 volts (30-40 electrolytic cells) he described "une secousse dans la tête" and, after recovering from that, the sound of

bubbling thick soup.[8] Rather than being put off by this, a certain Mr. Ritter, a contemporary of Mr. Volta, repeated the experiment on himself, only this time using 100-200 cells. This at the very least constituted a dangerous undertaking and the description of its effects probably kept other researchers from further experimentation for many years.[9] Another such experiment was conducted in 1855 by Duchenne of Bologna using alternating rather than direct current. This also did not produce a natural sound ("the sound of a fly between a curtain and a window") (cited in Simmons[10]). This phenomenon was investigated deeply in 1868 by Brenner, who varied such factors as polarity, frequency and intensity of the current and also used a form of bipolar stimulation of the ear. Brenner was able to generate a more natural sound with less unpleasant side effects (cited by Simmons[10]). Stevens and Jones (1939) showed that various mechanisms could be responsible for the generation of sound by electrical stimulation. Firstly, the middle ear can function as an electric transducer, thus being caused to vibrate by an electric current applied at the site of the outer ear canal , creating a sensation of sound (the "cochlear electrophonic" phenomenon). The basilar membrane of the cochlea can also act as such a transducer, with sound being generated in people lacking a middle ear structure. Finally, it also proved possible to generate a sensation of hearing in completely deaf people which has been attributed to direct stimulation of the auditory nerve. Djourno and Eyries (1957) conducted further clinical research by electrically stimulating the hearing nerve in a patient during an ear operation in connection with cholesteatoma.[11] Research in France demonstrated the possibility of perceiving sounds converted into electric current so that a few words could even be understood such as "papa," "maman," and "allo." Differences in current frequency are perceived as differences in tone pitch. Doyle was the first to attempt to make use of the cochlea's tonotopic features by placing a number of electrodes more or less arbitrarily in the cochlea of a patient with full sensorineural deafness. This made it possible for the patient in question to understand parts of sentences. Simmons et al.[10, 12] placed six electrodes along the medial wall of the cochlea (the modiolus) intending to stimulate various frequency areas of the nerve separately and simultaneously. It indeed proved to be the case that identical electric stimulation in various places of the nerve incited various tone pitches.

The clinical applicability of cochlear implantation got underway through the work of pioneers such as otologist William House, the brothers John and Jim Doyle, Jack Urban, Robin Michelson, Karen Berliner, the Australian Graeme Clark, and many others. House, Berliner and the Doyles laid the foundations for the clinical application of single-channel cochlear implants with one active and one reference electrode by producing the first experimental clinical cochlear implant in the world in 1962.[13] At the same time, the Australian group Clark and Tong were focusing on the development and application of a multichannel system (the Nucleus multichannel cochlear implant).[14] Other systems also came on the market, the most important of which came from Vienna (the 3M and later

the Med-El prosthesis by Hochmair & Burian) and Salt Lake City (the Ineraid prosthesis by Eddington produced by Symbion).

Since then the development of the cochlear implant has gained momentum. In 1984 the single-channel 3M/House implant was officially approved as the first sense-replacing apparatus by the US Food and Drug Administration (FDA), with the Nucleus multichannel system following a year later. Other systems that have been developed are the MXM system by Digisonic, the Clarion prosthesis by Advanced Bionics and the Laura implant developed in Antwerp. As has been the case everywhere in the electronics industry, significant miniaturization has taken place improving the applicability of CI in children.

New insights have also arisen and important developments have taken place regarding the design and placement of electrodes in the cochlea. For example, a number of systems now implement a form of so-called "modiolar hugging electrode arrays" with electrodes brought closer to the medial wall of the scala tympani and thus to the spiral ganglion cells. Experimentation is also being done with directional microphones to improve speech understanding in noisy situations.[15,16] The most progress in technology has probably taken place in the method of sound coding by the speech processor. This development was only made possible after provisional clinical implant programs had prompted a new round of fundamental research into the functioning of the auditive system.

Many studies have since the clinical application of CI, demonstrated the positive effect of CI on acoustic functioning.[17] Besides improved sound perception also the quality of speech production increases due to CI.[18] The stigmatizing effect of "deaf-speech" serves as a warning against underestimating this effect. Additional attention has been paid in recent years to the overall effect on quality-of-life including psychological self-esteem and social functioning. Not being able to hear not only impedes communication but can also serve to exclude people in countless areas. CI can contribute to the sense of belonging among such people. This dissertation pays ample attention to such quality of life aspects.

The number of deaf people rehabilitated with the help of CI has increased exponentially in recent years. In 1991 this number came to 4000 worldwide; now the number of Nucleus implants alone exceeds 26,000.[19] Since the FDA approved of the application of the Nucleus system in children in 1990, more than 10,000 children have obtained CI. Cochlear implantation has grown in recent years into a globally applied treatment of total deafness that no longer can be ignored.

ANATOMY AND PHYSIOLOGY

The ear can be divided into three parts (Figure 1): the outer portion comprising the auricle and external ear canal; the middle ear, comprising the membrana tympani and the space behind it containing the ossicles; and finally the inner ear, comprising the organ of

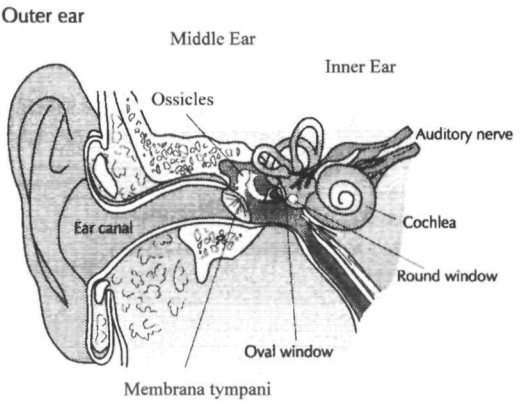

Figure 1. Cross-section of the ear (courtesy Med-El)

equilibrium and the auditory organ, i.e., the organ of Corti in the cochlea. The cochlea comprises a tube in the form of a snail's shell rolled up about 2 ½ times, in length divided into three smaller tubes, the scala vestibuli, the scala media and the scala tympani.

Sounds received by the ear travel through the ear canal and cause the membrana tympani to vibrate. The first ossicle, the malleus, is partially subsumed in the ear drum and transfers the vibration via the incus to the stapes. The stapes is connected to the cochlea through the oval window; it causes the liquid in the cochlea and thence, the basilar membrane to vibrate. The vibrations of the basilar membrane cause transformations in the hair cells of the organ of Corti, generating nerve impulses. The perception of tone pitch is organized in the cochlea in a so called "tonotopic" manner in which high pitched tones are relayed by the basal parts of the organ of Corti and the low pitch tones in the more apical parts. The nerve impulses are passed on by dendrites to the ganglion spirale, consisting of the cell bodies of these dendrites. Impulses run from the ganglion to the cochlear nuclei in the brain stem and further up the auditory pathway.

Diseases of the auricle, ear canal and the middle ear can lead to a reduction in hearing (conductive hearing loss) but not to total deafness. Total sensorineural deafness is always the result of serious damage to the cochlea, auditory nerve or parts of the central auditory system (brain stem, cerebrum). Damage to the cochlea is by far the most common cause of sensorineural hearing loss (for example, presbyacusis); diseases of the neural system are only rarely the cause of total deafness.[20] Cochlear implantation is not possible in such cases.

Despite so called "retrograde degeneration" of nerve fibers extending from the hair cells to the ganglion spirale, many of the cell bodies in the ganglion spirale remain intact, as do their axons that run to the cochlear nuclei in the brain stem. The number of surviving ganglion cells is partly determined by the etiology of the deafness: the highest survival rate is found with sudden idiopathic deafness, while the lowest level is found with post-viral labyrinthitis, congenital or hereditary deafness or bacterial meningitis.[21,22,23,24,25,26] The survival of a significant portion of the auditory system makes it possible to stimulate a sufficient amount of nerve cells of the auditory nerve via electrodes in a "deaf cochlea." This is illustrated by the fact that nearly all deaf candidates for CI have auditory sensations

when submitted to a pre-operative promontory stimulation test.[27]

The incidence of clinically significant hearing loss in the general population varies from 10% to 20% depending on the exact definition of hearing loss.[28] No reliable data are available regarding the exact number of persons suffering from total sensorineural deafness in the Netherlands. According to the Central Bureau for Statistics, 0.2% of the Dutch population suffers from an extremely serious degree of hearing restriction.[29] Extremely serious hearing restriction was defined as the inability to hear with an adequate conventional hearing aid loud noises such as a car horn or what is being said in a conversation with one person. This percentage lines up well with an earlier estimate of 0.21% made by Thornton for serious hardness of hearing, defined precisely by him as an average hearing loss greater than 90 dBHL in the best ear.[30] In the Dutch situation (15 million inhabitants) this would equal 31,000 inhabitants including children and pre- and post-lingual deaf adults. According to Thornton, 0.007% of the adult population (18-65 years) is post-lingually deaf (average hearing loss of 110 dBHL or more). Looking at adults between 18 and 75 years of age, the number of adults in the Netherlands who would come under consideration for a cochlear implant would equal 1400 persons. Experience from the Nijmegen implant program teaches that some 30% of these persons satisfy all selection criteria for CI, yielding about 420 people actually being considered for implantation. Due to the technological improvements that have been made in the field of CI, people with some remaining capacity for hearing, who until recently would have been rehabilitated with a conventional hearing aid, would now be better off with a CI.[31] Expanding the hearing selection criterion to for example an average hearing loss of 100 dBHL instead of 110 dBHL would bring an extra 1360 persons under consideration for cochlear implantation.

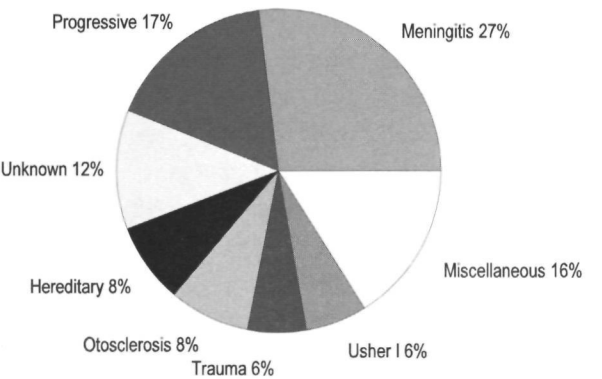

Figure 2. Etiology of deafness in CI adult users treated in Nijmegen.

Causes of deafness in adult CI users treated in Nijmegen are shown in Figure 2.

THE COCHLEAR IMPLANT

The current generation of implant systems comprises an externally worn part and an operatively placed internal part. The external part comprises a small microphone behind the ear connected to a speech or sound processor that codes the received sounds. Speech processors are body worn or, increasingly, behind the ear as well. The coded information is sent via a radio frequency transmitter to the receiver portion of the implant located under the skin. The transmitter is magnetically linked to the subcutaneously located receiver portion. The receiver portion subsequently emits electric signals to electrodes located in the cochlea (Figures 3 and 4).

In the later models a back-telemetry link provides information concerning the electrical integrity of the implant. Much research has been invested into the best modes of sound or speech coding. The early single-channel implants used an analog form of signal processing. The speech signal was amplified, filtered and emitted to an electrode placed in the cochlea or at the site of the round window. Although the system was successfully implanted in many patients, it has shown its limitations[32,33] nor can the single-channel implant make use of the tonotopic structure of the cochlea since the tuning curves of all fibres are essentially flat.

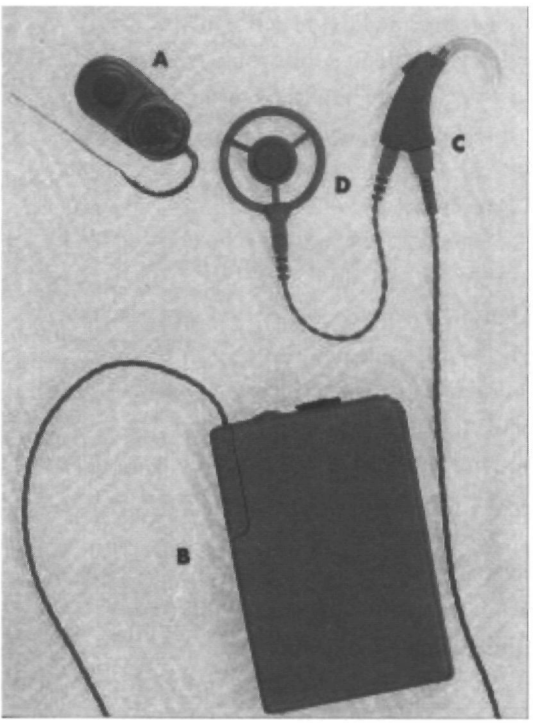

Figure 3. Different parts of the Nucleus 22 channel implant.
A = internal part consisting of receiver and electrode casing;
B = speech processor (MSP);
C = behind-the-ear part with microphone;
D = transmitter coil.

With a multichannel implant the electrodes are placed intra-cochlearly along the basilar membrane. The first most widely used system with this approach was the Nucleus 22 channel system made by the Cochlear Corporation. Several speech processing strategies were developed for this system over the years.[34] The first was applied in the Wearable Speech Processor (WSP).[35] This implemented a formant extraction strategy which attempted to extract the most important aspects from the speech signal, viz the ground tone F0 and second formant F2. F0 was represented as stimulus repetition rate, F2 by stimulating one of the electrodes depending

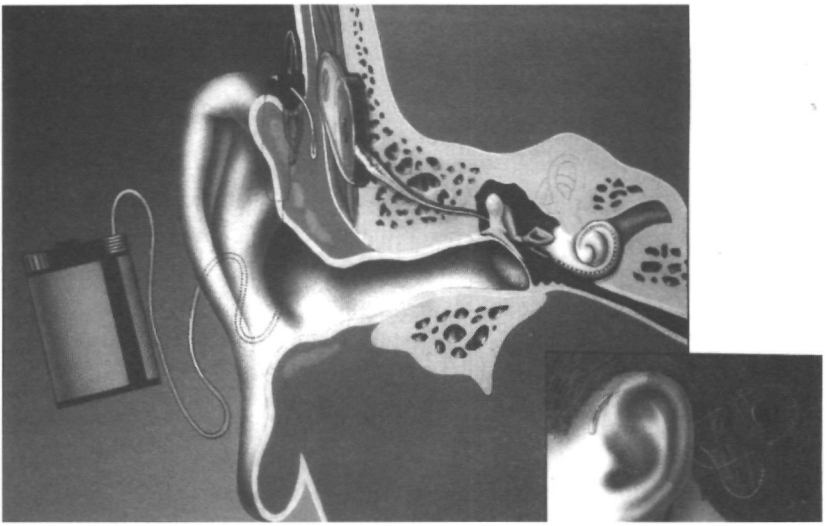

Figure 4. The various external and internal parts of a multichannel cochlear implant system (courtesy Cochlear Ltd.)

on its frequency. The first formant, F1, was added later in an upgrade of the WSP. Now the 22 channels were divided into two groups, one for F1 and one for F2. The Mini Speech Processor (MSP) added filters aimed at extracting the third, fourth, and fifth formats. These formants were assigned a fixed set of electrodes in the basal part of the electrode array. to the coded signal because these also play an important role in the understanding of consonants. By directing various electrode combinations, various speech sounds can be transmitted in this manner. This technique was called the Multipeak strategy (MPEAK)[36] (shown in Figure 4) and demonstrated improved results on speech perception tests.[37] Most of the implanted subjects described in this dissertation were fitted with the Nucleus 22 channel implant and the MSP. The disadvantage of this method is that speech cannot be easily distinguished from background noise and certainly not from other competing speech sounds. The Spectral Peak (SPEAK) coding developed later was intended to be more effective in this regard.[38] In this strategy up to ten peaks in the spectrum rather than the limited number of peaks corresponding to the 5 formants are extracted from the speech signal. This improved speech perception especially in a noisy environment.[39,40]

The Continuous Interleaved Sampling (CIS) strategy was developed with the same intent to convey spectral information in a robust manner.[40,41] As SPEAK, the CIS strategy does not derive certain features from the speech signal. The entire signal is bandpass filtered in as many bands as there are electrodes and each electrode is stimulated at a fixed rate way above F0 (that is about 800 pulses per second per electrode). This is effectuated by high speed electrode activation with short pulse signals and by restricting the number

of electrodes to 8-12. There are studies indicating that even the best performing implant users would not benefit from increasing the number of electrodes beyond 8 to 12, at least for understanding speech in quiet.[42] Most implant manufacturers have implemented this strategy or are working on it in view of the results that have been achieved.[43,44] The lately more widely used eight-channel Clarion system made by the Advanced Bionics Corporation is equipped with the capability of simultaneous analog speech processing along with the CIS strategy. The analog speech signal is divided among eight band filters and immediately transmitted to the various corresponding electrodes. The basic principle of this system is in fact closely related to the analog system applied in the first single-channel implants, the major difference being that it presents the signal via eight electrodes located in various frequency specific places in the cochlea rather than via only one electrode. One important other difference is the ability to stimulate between two adjacent electrodes in the cochlea ("bipolar mode"), which minimizes the overlap of the electrical fields, instead of stimulating between one intra and one extra-cochlear electrode (monopolar mode) as in the older devices. It is claimed that the new "modiolar hugging" electrode arrays even further improve the channel-separation to increase the benefit of simultaneous stimulation. It is however unclear to what extent the analog signal is superior to the (high rate) pulsatile stimulus used in other strategies. Eventually it is hoped that stimulation will be possible at many clearly separated sites along the basilar membrane with an excitation signal that closely matches the (temporal and dynamic) requirements of the local nerve populations.

THE SITUATION IN THE NETHERLANDS

In 1985 the first cochlear implant surgery in the Netherlands was performed in Utrecht. The Nijmegen Cochlear Implant program started in 1987. The first favourable experiences led to a request to the health authorities to reimburse the treatment. Before they would make a decision on this matter, however, two health-care development projects being financed by the national health insurance program had to be completed first. The results of the first project, carried out by the Nijmegen University Hospital in cooperation with the Sint Michielsgestel Institute for the Deaf, were published in the report "Cochlear Implants."[45]

This report listed the selection procedure, the surgical technique, the device characteristics, rehabilitation and results gained in this period regarding the auditive functioning of 20 implanted adults. Results were favorable and received further support from research on a much larger scale abroad; nevertheless, the so-called electric inner ear prosthesis failed to gain inclusion in health insurance coverage. However, the second project was financed providing for the implantation of another 40 postlingual deaf adults in a joint venture between the university hospitals of Utrecht and Nijmegen and the Sint

Michielsgestel Institute for the Deaf. This project was intended to define the selection criteria for CI more precisely, to demonstrate its effect on quality of life and to obtain a better idea of costs involved. The result was a second report entitled "The Selection of Deaf Adults for an Electric Inner Ear Prosthesis (Cochlear Implant) and the Evaluation of Results Obtained with This Treatment" (October 1991-October 1994).[46]

The report formulated clear selection criteria (see below), confirmed favorable effects on communication and contact with surroundings and made plausible CI's positive impact on the quality of life. It also made recommendations regarding the number of implants expected per year, the protocol to be followed, the organizational structure in the Netherlands and required post-operative care.

A third health-care development project was begun, inspired by favorable results of CI in children at home and abroad. The project involved cochlear implantation in 20 children. Timely measures to compensate deafness are essential especially in elected children because their speech development is still in full progress; by this time so much was known about the favorable effect of CI on speech development that the treatment could no longer be withheld from them on the grounds of insufficient experience with CI in children. Results were described in the report "Cochlear Implantation in Children" (March 1993-March 1996).[47]

As was the case with the studies dealing with adults, the report was in line with findings of large foreign studies.[48 49 50]

Although the clinical application of cochlear implantation has been restricted in the Netherlands to the university hospitals in Utrecht and Nijmegen, theoretical and experimental research into cochlear implants conducted at the University Hospital Leyden deserves mention.[51] Furthermore, other hospitals have shown interest in a CI program should funding be made available to them. Cochlear implantation for adults is an officially accepted treatment but thus far has only recently received a limited amount of funding resulting in long waiting lists. Cochlear implantation in children has recently been approved as a regular health provision for a limited number of patients. More research will be required into long-term results and the impact on the social and emotional development of children with a cochlear implant.

THE NIJMEGEN-SINT MICHIELSGESTEL PROTOCOL

The latest report from the national health insurance council regarding CI in adults devoted ample attention to the protocol to be followed upon reporting a candidate for CI. The following presents a resume from this protocol.

After application via a questionnaire, the selection process begins with an interview with an ENT doctor and extended audiometrical evaluation, including a hearing-aid trial if necessary. If no contra-indications are noted, this is followed by an image-forming

investigation of the petrous bone, possible test stimulation of the auditory nerve, evaluation of the vestibular function and an investigation by a social worker and/or psychologist.

The following inclusion criteria have been formulated:

1) total sensorineural deafness, i.e. no functional hearing remnant, not even with a hearing aid (Initially this came down to a PTA greater than 110 dB);
2) sufficient communicative and/or intellectual possibilities to make reasonable progress with rehabilitation;
3) realistic expectations and good motivation;
4) a partner (co-therapist) who can help with daily exercise at home;
5) perception of sound sensations in a promontory test;
6) an open scala tympani allowing electrodes to be placed in the cochlea;
7) normal anatomic relations and the absence of infections in the mastoid;
8) good general health.

Although these criteria remain valid in general, most conditions have been relaxed or exceptions to these rules have been made over the years. The audiometric criterion has shifted; nowadays also deaf people with some residual functional hearing are considered for CI. The daily need for a co-therapist has become less important with the introduction of more advanced implant systems leading to a shorter and mostly easier rehabilitation period. The condition requiring the open scala tympani has also been relaxed, as it has been demonstrated that a favorable result can be obtained even with an incomplete insertion of electrodes in the cochlea, although results are usually not as good as with a complete placement.[52,53]

Furthermore a number of tests are done (conventional speech audiometry as well as the closed-set "Antwerp-Nijmegen" tests) to assess the auditory capabilities before implantation. The same tests are also used to evaluate the progress and the results achieved after implantation.

After termination of the selection procedure a final decision is taken about implantation.

The operation requires a hospital stay of a few days. Upon release, post-operative wound healing is checked in the outpatient clinic.

A recovery period of about six weeks is followed by the initial programming of the processor. This is followed by hearing training, which in the case of an operation in Nijmegen takes place at the rehabilitation department of the Institute for the Deaf in Sint Michielsgestel over a period of some days. Preferably a couple of implant recipients is rehabilitated together with their co-therapists. After this, further rehabilitation finds place in the home accompanied by outpatient clinic processor attunements.

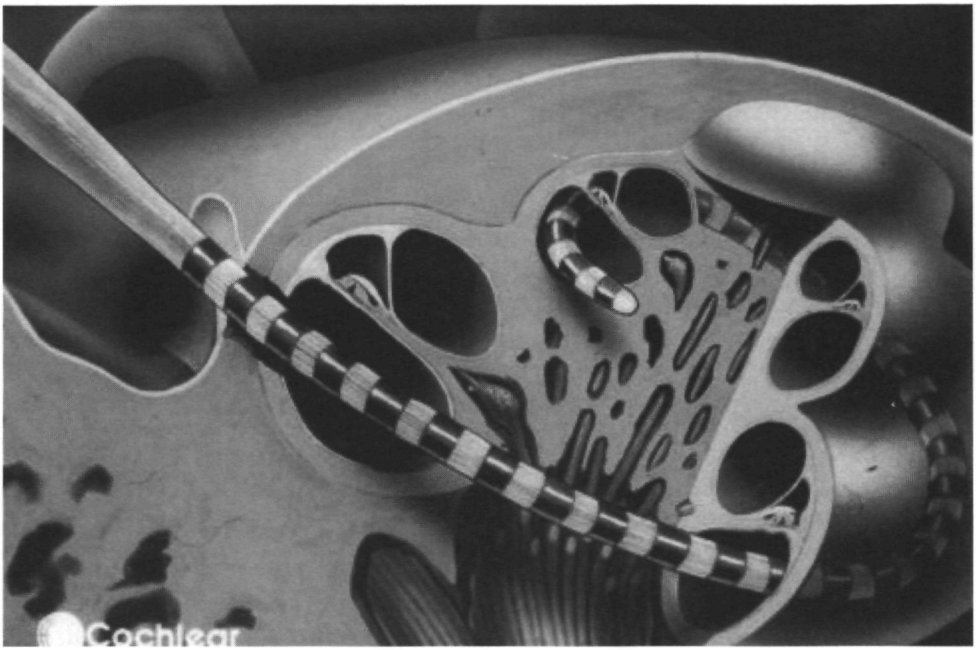

Figure 5. Nucleus 22-channel electrode-array positioned in the scala tympani of the cochlea (courtesy Cochlear Ltd)

Speech perception and other tests are conducted after three and 12 months and once every 2 years thereafter to evaluate the level of auditive functioning. A number of other aspects are periodically evaluated via questionnaires.

OPERATIVE PROCEDURE

Cochlear implant surgery is performed under general anesthesia via a posterior tympanotomy. This is a routine ear surgery technique used mainly to remove chronic ear infections. It involves drilling an entry into the middle ear through the mastoid. This entry provides a view of the round window. After opening this window, electrodes can be placed in the cochlea (Figure 5) and the receptor unit of the implant can be attached to the skull bone under the skin.

The correct placement of electrodes is checked via a conventional x-ray of the skull (Figure 6).

As with most surgical interventions, cochlear implantation involves risks. Besides general risks involved with general anesthesia, specific risks arise depending upon the implant type used, individual anatomy, possible pathology of the subject ear and the surgical technique used. Life-threatening complications have not been reported with cochlear implants.

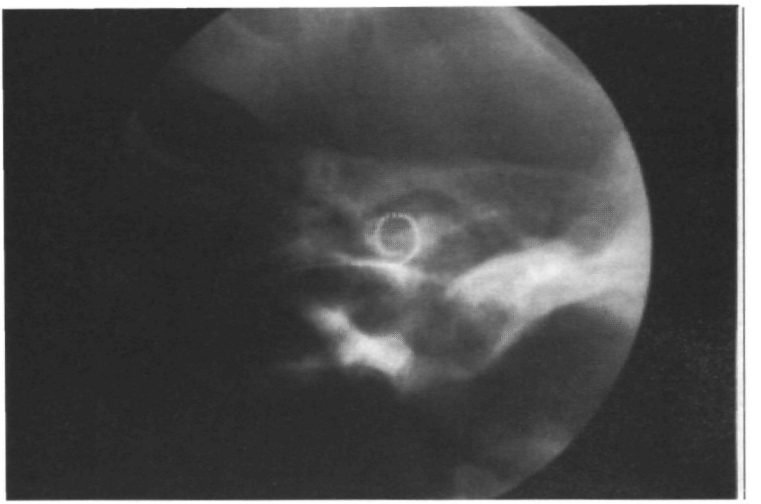

Figure 6. Stenvers projection conventional x-ray showing the correct placement of the electrode array in the basal turn of the cochlea

Significant complications are complications requiring corrective surgery such as faulty placement, problems with the skin flap, implant migration or rejection. Damage to the facial nerve is also viewed as a significant complication. Good surgical technique has however made such complications extremely rare. Significant complications arise in some 5% of cases of implants in adults.[54,55] In children this percentage is supposed to be lower. This is a low complication rate in comparison with, for example, the 10% rate from pacemaker implants.[56]

Furthermore, a number of minor complications have been distinguished not requiring surgical intervention. The most prevalent is probably the undesired stimulation of the facial nerve when the implant is activated by sound. Kelsall reports the prevalence of 7%, the largest portion of which found in conjunction with otosclerosis. The usual treatment is adjusting the attunement of the processor.[57] Tinnitus is also reported as a complication, but just as many cases report a favorable effect on previously existing tinnitus after CI.[58] Short-lived dizziness has also been reported after implantation in one ear in which the vestibular system is still functional.[59] A final minor complication to be noted involved the non-functioning of a few specific electrodes to which the processor can also be attuned if properly diagnosed.[60]

In the Nijmegen situation there have been nine cases of defective or poorly functioning implants over the years (3 single-channel implants, one Nucleus implant, 5 other systems: Med-El and Laura) requiring re-implantation. Re-implantation required by faulty placement of electrodes was performed in one case while another in another case the implant needed to be replaced in connection with facial nerve stimulation. In the case of the Nucleus system – the most used worldwide – the report after nine years is that 95% of

the implants still function properly.[61,62]

EVALUATION OF ACOUSTIC FUNCTIONING

Evaluation of the effect of the implant is obtained via free field tone and speech audiometry. Special tests have been developed for CI in order to evaluate various levels of sound and speech perception. Tests have also been developed to measure lip reading ability. The tests described in this dissertation stem from the Antwerp/Nijmegen test battery (AN tests).[63] This comprises a number of tests that measure auditive capabilities regarding environmental sounds and different levels of speech perception.

In addition, lipreading ability is measured both with and without an implant with the help of the Connected Discourse Tracking method (CDT). the researcher reads a story line by line. The number of words the test person can repeat per minute is then measured. This number is compared with the speed at which the subject reads a similar text aloud (the top score).[64]

RESULTS

The 1991 and 1995 reports for the National Health Insurance Council regarding the two development health care projects treating deaf adults indicated that acoustic contact with the outside world is restored in nearly all implant users. Of these, 61% are in a condition to understand speech conducted at a normal rate in combination with lip reading. After 12 months of use, 26% of a randomly selected group of speech sounds are understood without lip reading (until 1994 mostly with the Nucleus implant with the MSP system). These percentages have since improved thanks to improved speech coding strategies as mentioned earlier. Staller et al. report that within a half-year of implant use 43% of implant recipients attain over 90% correct recognition of everyday sentence without lip reading.[65] Implant users' speech becomes easier to understand and more natural. The implant also appeared to contribute to the patient' s psychological and social functioning and self-esteem.[45,46]

AIM OF THE STUDY

Research in the area of cochlear implants is motivated by the desire to restore hearing as much as possible while causing minimum side effects. This has prompted research across a wide range of areas. Firstly, fundamental knowledge is required into the mechanisms by which the normally intact auditory system works as well as into the pathological mechanisms in deafness. Diagnostic tools are necessary to gather as much relevant information as possible in the pre-operative stage to minimize the risk of undesired

surprises such as cochlear obstructions or post-operative dizziness or the possibility of a non-functioning auditory nerve and so on. Technological research into electronics, biomechanics and biosafety is also important to the production of optimal hearing prostheses. Surgical techniques have to be developed and sophisticated to make the surgical procedure safe and effective. Finally, clinical research provides the standard by which all else is measured; in the final quality-of-life analysis, success is determined by the degree to which the user of the hearing prosthesis actually benefits.

The Nijmegen-Sint Michielsgestel Cochlear Implant program started in 1987 with the implantation of a single-channel implant in a postlingually deafened adult. Since then around 200 deaf individuals have undergone cochlear implantation in our centre including 100 children. Most of these implantations have been done as part of research projects evaluating many aspects of cochlear implantation.

This dissertation focuses on some specific aspects of risk reduction and less frequently reported results in cochlear implantation.

1. An important aspect of the preoperative assessment is to know the shape and patency of the cochlea as judged by radiological examinations like Computer Tomography (CT) and Magnetic Resonance Imaging (MRI). The exact place and value of these examinations is still being debated, notwithstanding the agreement that it is of prime importance to have as accurate information as possible before surgery. Until recently, CT scans provided the most precise images of the mastoid and the cochlea. Although CT gives extremely detailed recordings of the bony structures of the ear, non-osseous irregularities such as fibrous obstructions of the cochlea are not detected. With the perfecting of MRI technology, it has recently become possible to view the contents of the cochlea and identify osseous and non-osseous obstructions as an interruption of the normal signal from the cochlea. As part of the development health care projects, every CI candidate received a CT scan of both ears; the last project also included MRI scans for adults. Results are compared with operative findings and discussed.
2. Cochlear implantation involves opening the inner ear (scala tympani) and therefore runs the risk of damage to the inner ear structures including the vestibular labyrinth. Little is known about the possibility of damage to the vestibular function. In the Nijmegen-St Michielsgestel protocol, vestibular functioning of both labyrinths was evaluated in every patient pre- and post-operatively to assess this risk.
3. Cochlear implantation in prelingually deafened adults has been controversial from the start because of the limited benefit expected in this group. Yet experience has taught that truly auditory percepts are evoked, even in congenitally deaf individuals, and in some patients the benefit may be crucial for their functioning in society.[66] This is especially true for patients with Usher syndrome, a form of hereditary

deafness accompanied by a progressive form of retinitis pigmentosa, an affliction to the eye that eventually leads to a serious reduction of vision, ultimately even blindness (Usher type I syndrome).[6]

The results of several adult patients suffering from Usher syndrome type I are compared with those of other prelingually deafened individuals and with postlingually deafened individuals.

4. New health care provisions are nowadays increasingly subject to close scrutiny especially with regard to quality of life. Until recently, only sound and speech perception data were presented as a measure of the success of cochlear implantation, with a sprinkling of subjective findings from qualitative questionnaires and interviews with patients, presented without structure. In view of the rapidly rising costs in health care, the need is increasingly being felt for information regarding the consequences of improved sound and speech perception in relation to cost-benefit analysis. This trend is also observable in other medical interventions. It is also of interest to policymakers to be able in this manner to compare various medical interventions. This dissertation describes the development of a Health-Related-Quality-of-Life (HRQoL) questionnaire for CI: the Nijmegen Cochlear Implant Questionnaire (NCIQ) and its application.

   The results of the NCIQ were also closely analyzed and compared with two other generic quality of life questionnaires (i.e., not developed for any specific affliction), the Short-Form 36 and the Health Utilities Index, within the framework of the same investigation.

   Finally the results of the various studies included in this dissertation are summarized and discussed.

REFERENCES

1. Brooks PL, Frost BJ, Evaluation of a tactile vocoder for word recognition, J Acoust Soc Am 1983;74:34-39
2. Cohen NL, Waltzman SB, Fischer SG. A prospective randomized study of cochlear implants. New Eng J Med 1993;328:233-237
3. Waltzman SB, Fisher SG, Niparko JK, Cohen NL. Predictors of postoperative performance with cochlear implants. Ann Otol Rhinol Laryngol (suppl) 1995;165:15-18
4. Blamey P, Arndt P, Bergeron F, Bredberg G, Brimacombe J, Facer G, Larky J, Lindström B, Nedzelski J, Peterson A, Shipp D, Staller D, Whitford L. Factors affecting auditory performance of postlingually deaf adults using cochlear implants. Audiol Neurootol 1996;1:293-306
5. Tyler RS, Parkinson AJ, Woodwordth GG, Lowder MW, Ganz BJ. Performance over time of adult patients using the Ineraid or Nucleus cochlear implant. J Acoust Soc Am 1997;102:508-522
6. van Aarem, A. Heterogeneity in the Usher Syndrome. Thesis, University of Nijmegen, 1996
7. Clark GM. Middle ear and neural mechanisms in hearing and in the management of deafness. Thesis, University of Sidney, 1969
8. Volta A. On the electricity excited by mere contact of conducting substances of different kinds. Trans Royal Soc Philos 1800;90:403-431 cited by Simmons FB, 1966
9. Ritter JW. Wirkung des Galvanismus der Voltaischen Batterie auf menschliche Sinneswerkzeuge, Annal Physsik 1801;7:447-484 cited by Simmons FB, 1966
10. Simmons FB. Electrical stimulation of the auditory nerve in man. Arch Otolaryngol 1966;84:2-54
11. Djourno A, Eyries C. Prosthese auditive par excitation electrique a distance du nerf sensoriel a l'aide d'un bobinage inclus a demeure. Presse Medicale 1957;35:14-17
12. Simmons FB, Epley JM, Lummis RC. Auditory nerve: electrical stimulation in man. Science 1965;148:104-106
13. House LR. Cochlear Implant: The beginning, Laryngoscope 1987;97:996-997
14. Clark GM, Tong YC, Black R, Forster IC, Patrick JF and Dewhurst DJ: A multiple-electrode cochlear implant. J Laryngol Otol 1977;91:935-945
15. Wilson BS. The future of cochlear implants (editorial). Br J Audiol 1997;31:205-225
16. Clark GM. Research advances for cochlear implants. Auris Nasus Larynx 1998;25:73-87
17. Makhdoum MJ. Auditory performance of cochlear implant users-psychophysical and objective measurements. Thesis, University of Nijmegen, 1998
18. Langereis MC. Effects of cochlear implantation on speech production. Thesis, University of Utrecht, 1997
19. Cochlear Ltd web site (http://www.cochlear.com)
20. van Leeuwen JPPM. Acoustic neuromas. Diagnostic aspects and results of surgery. Thesis, University of Nijmegen, 1995
21. Kerr AG, Schuknecht HF. The spiral ganglion in profound deafness. Acta Otolaryngol (Stockh) 1986; 65:586-598
22. Hinojosa R, Marion M. Histopathology of profound sensorineural deafness. In: Parkins CW, Anderson SW, eds. Cochlear prosthesis: an international symposium. Ann N Y Acad Sci 1983;405:459-484
23. Suzaka Y, Schuknecht HF. Retrograde cochlear neuronal degeneration in human subjects. Acta

Otolaryngol (Stockh) 1988 Suppl 450

24 Nadol JB, Young YS, Glynn RJ Survival of spiral ganglion cells in profound sensorineural hearing loss implications for cochlear implantation Ann Otol Rhinol Laryngol 1989,98 411-416

25 Nadol JB Degeneration of cochlear neurons as seen in the spiral ganglion of man Hear Res 1990,49 141-154

26 Nadol JB Patterns of neural degeneration in the human cochlea and auditory nerve implications for cochlear implantation Otolaryngol-Head-Neck-Surg 1997,117 220-228

27 Spies TH, Snik AFM, Mens LHM, van den Broek P Preoperative electrical stimulation for cochlear implant selection Acta Otolaryngol (Stockh) 1993,113 579-584

28 Grote JJ Slechthorendheid, een vergeten volksgezondheidsprobleem Ned Tijdschr Geneeskd 1992,136 2404-2406 (In Dutch)

29 Centraal Bureau voor Statistiek, Lichamelijke beperkingen bij de Nederlandse bevolking 1986/1988, 's-Gravenhage, SDU-Uitgeverij/CBS-publicaties, 1990 (In Dutch)

30 Thornton, AR Estimation of the number of patients who might be suitable for cochlear implant and similar procedures British Journal of Audiology 1986,20 221-229

31 Muller-Deile J, Rundert H, Brademan G, Frese K Cochlear Implant Versorgung bei nicht tauben Patienten? Laryngorhinootologie 1998,77 136-143

32 Clark GM A neurophysiological assessment of the surgical treatment of perceptive deafness Int Audiol 1970, 9 103-109

33 Eyles JA, Aleksy WL, Boyle PJ Performance changes in University College Hospital/Royal National Institute for the deaf single-channel cochlear implant users upgraded to the Nucleus 22-channel cochlear implant system Ann Otol Rhinol Laryngol (suppl) 1995,166 263-265

34 Mens LHM, Brokx JPL, van der Harten JJM, Hinderink JB, Peters-Bos M, Vermeulen-van Werde L, van den Broek P Auditieve waarneming bij dove volwassenen met een cochleair implantaat Stem-, Spraak-, en Taalpathologie 1996,5 31-44 (In Dutch)

35 Tong YC, Clark GM, Dowell RC, Martin LFA, Seligman PM, Patrick JF A multiple-channel cochlear implant and wearable speech-processor An audiological evaluation Acta Otolaryngol 1981,92 193-198

36 Koch DB, Seligman PM, Daly C, Whitford LA A multipeak feature extraction coding strategy for a multichannel cochlear implant Hear Instr 1990,41 28-30

37 Cohen NL, Waltzman SB Influence of processing strategies on cochlear implant performance Ann Otol Rhinol Laryngol 1995,165 9-14

38 Skinner MW, Clark GM, Whitford LA, Seligman PM, Staller SJ, Shipp DB, Shallop JK, Everingham C, Menapace CM, Arndt PL Evaluation of a new spectral peak (SPEAK) coding strategy for the Nucleus 22 Channel Cochlear Implant System Am J Otol (suppl) 1994,15 15-27

39 Holden LK, Skinner MW, Holden TA Speech recognition with the MPEAK and SPEAK speech-coding strategies of the Nucleus cochlear implant Otolaryngol Head Neck Surg 1997,116 163-167

40 Wilson BS, Finley CC, Lawson DT, Wolford RD, Eddington DK, Rabinowitz WM Better speech recognition with cochlear implants Nature 1991,352 236-238

41 Wilson BS, Finley CC, Lawson DT, Wolford RD, Zerbi M Design and evaluation of a continuous interleaved sampling (CIS) processing strategy for multichannel cochlear implants J Rehab Res Develop 1993,30 110-116

42 Brill SM, Gstottner W, Helms J, Von Ilberg C, Baumgartner W, Muller J, Kiefer J Optimization of channel number and stimulation rate for the fast continous interleaved sampling strategy in the

Combi 40+ Am J Otol (suppl) 1997,18 104-106
43 Boex C, Pelizzonne M, Montadon P Speech recognition with a CIS strategy for the Ineraid multichannel cochlear implant Am J Otol 1996,17 61 68
44 Kiefer J, Muller J, Pfenningdorff T, Schon F, Helms J, Von Ilberg C, Baumgartner W, Gstottner W, Ehrenberger K, Arnold W, Stephan K, Thumfart W, Baur S Speech understanding in quiet and noise with the CIS speech coding strategy (Med-El Combi-40) compared to the multipeak and spectral peak strategies (Nucleus) ORL-J-Otolaryngol-Relat-Spec 1996,58 127-135
45 Van den Broek P, et al Cochlear Implants Main report 1991 University Hospital Nijmegen (In Dutch)
46 Van den Broek P, et al Selectie van volwassen doven voor een elektrische binnenoorprothese (cochlear implant) en evaluatie van de met deze prothese verkregen resultaten Main report 1995 University Hospital Nijmegen (In Dutch)
47 Van den Broek P, et al Cochleaire Implantatie bij kinderen Main report 1996 University Hospital Nijmegen (In Dutch)
48 Cochlear Implants in Adults and Children NIH Consensus Statement 1995 May 15-17,13 1-3
49 Miyamoto RT, Svirsky M, Kirk KI, Robbins AM, Todd S, Riley A Speech intelligibility of children with multichannel cochlear implants Ann Otol Rhinol Laryngol (suppl) 1997,168 35-36
50 Waltzman S, Cohen NL, Gamolin R, Green J, Shapiro W, Brackett D, Zara C Speech perception and production results in children implanted between 2 and 5 years of age Adv Otolaryngol 1997,52 177-180
51 Frijns JHM Cochlear Implants A modelling approach Thesis, University of Leyden, 1995
52 Cohen NL, Waltzman SB Partial insertion of the Nucleus multichannel cochlear implant technique and results Am J Otol 1993,14 357-361
53 Kirk KI, Sehgul M, Miyamoto RT Speech perception performance of Nucleus multichannel cochlear implant users with partial electrode insertions Ear Hear 1997,18 456-471
54 Cohen NL, Hoffman RA, Strischein M Medical or surgical complications related to the Nucleus multichannel cochlear implant Ann Otol Rhinol Laryngol 1988,97 8-13
55 Windmill IM, Martinez SA, Nolph MB, Eisenmenger BA Surgical and nonsurgical complications associated with cochlear prosthesis implantation Am J Otol 1990,11 415-420
56 Ferguson TB, Lindsay BD, Boineau JP Should surgeons still be implanting pacemakers? Ann Thorac Surg 1994,57 588-597 Brokx JPL, Beijnon AJ, Durme M van, Timmermans Ontwikkeling van Nederlandstalige auditieve tests Logopedie en Foniatrie 1987, 6 113 116 (In Dutch)
57 Kelsall DC, Shallop JK, Brammeier TG, Prenger EC Facial nerve stimulation after Nucleus 22-channel cochlear implantation Am J Otol 1997,18 336-341
58 Tyler RS, Advantages and disadvantages expected and reported by cochlear implant patients Am J Otolaryngol, Head Neck Surg 1994,15 523-531
59 Huygen PLM, Hinderink JB, van den Broek P, van den Borne S, Brokx PLM, Mens LHM, Admiraal RJC The risk of vestibular function loss after cochlear implantation Acta Otolaryngologica (Stockh) (suppl) 1995,520 270-272
60 Mens LH, Oostendorp T, van den Broek P Identifying electrode failures with cochlear implant generated surface potentials Ear Hear 1994,15 330 8 De Filippo CL, Scott BL A method for training and evaluating the reception of ongoing speech J Acoust Soc Am 1987, 63 1186-1192
61 West RE, Stucky J Cochlear Implantation Outcomes experience with the Nucleus 22 implant Ann Otol Rhinol Laryngol (suppl) 1995,166 447-9

62. von Wallenberg EL, Brinck JM. Cochlear Implant Reliability. Ann Otol Rhinol Laryngol (suppl) 1995;166:441-443
63. Brokx JPL, Beijnon AJ, Durme M van, Timmermans. Ontwikkeling van Nederlandstalige auditieve tests. Logopedie en Foniatrie 1987; 6:113-116 (In Dutch)
64. De Filippo CL, Scott BL. A method for training and evaluating the reception of ongoing speech. J. Acoust. Soc. Am 1987; 63:1186-1192
65. Staller S, Menapace C, Domico E, Mills E, Dowell RC, Geers A, Pijl S, Hasenstab S, Justus M, Brunelli T, Adam A, Borton T, Lemay M. Speech perception abilities of adult and pediatric Nucleus implant recipients using the Spectral Peak (SPEAK) coding strategy. Otolaryngol Head Neck Surg 1997;117:236-242
66. Zwolan TA, Kileney PR, Telian SA. Self-report of cochlear implant use and satisfaction by prelingually deafened adults. Ear Hear 1996;17:198-210

CHAPTER TWO

MRI AND HRCT IN THE PREOPERATIVE WORK-UP FOR COCHLEAR IMPLANTATION

J.B. Hinderink, MD, F.B.M. Joosten, MD, PhD,
P. van den Broek, MD, PhD

Submitted for publication

MRI AND HRCT IN THE PREOPERATIVE WORK-UP FOR COCHLEAR IMPLANTATION

J.B. Hinderink, MD, F.B.M. Joosten, MD, PhD,
P. van den Broek, MD, PhD

ABSTRACT

Purpose: To describe results and compare advantages and disadvantages of different imaging techniques for the preoperative radiological evaluation of cochlear implant candidates.
Methods: Radiological and surgical findings were compared in 100 consecutively implanted patients. All patients underwent high resolution CT scanning of both petrous bones; 28 also underwent MRI.
Results: Meningitis was the main cause of deafness (45/100) and the only etiology that caused clinically significant compromised cochlear patency (17/45). CT achieved a Sensitivity of 78% for detecting diffuse cochlear obstruction while MRI achieved 100%. Other abnormalities were detected equally well by CT and MRI imaging, although CT imaging provided more detailed information about bony structures. On the other hand, MRI provided additional information about the internal auditory canal and other possible retrocochlear abnormalities.
Conclusion: In cochlear implant candidates with an etiology of meningitis, consideration should be given to starting with MRI rather than CT scanning. The provision of more elaborate information about cochlear patency by MRI facilitates decision-making regarding whether to implant or not and, if so, what implant system to use. In patients with other causes of deafness, a CT scan is sufficient.

INTRODUCTION

In the past decade, high-resolution CT (HRCT) scanning has become a generally accepted routine in the preoperative work-up for cochlear implantation at most implant centres.[13,14] It not only provides information about specific anatomical abnormalities but, more importantly, it also informs the surgeon about compromised cochlear patency which might prevent the full insertion of intracochlear electrode arrays.

This is especially important in patients whose cause of deafness is labyrinthitis, otosclerosis or fractures of the bony labyrinth.[6,15] Although the positive predictive value of CT is reported to be good in this respect, several studies have reported a considerable number of false negative CT findings.[2,8,7,12,15] Such findings stimulated the use of MRI to predict the likelihood of the presence of fibrous obstructions in the cochlea or poorly mineralized ossifications undetectable by CT.[1,5,9,10] With the enormous improvements in the spatial resolution of MRI techniques in recent years,[3] the accuracy for assessing cochlear patency might now be expected to be superior to that of CT. Additionally, MRI provides highly detailed information about the cochlear and vestibular nerves and the central nervous system. Another obvious advantage of MRI is the lack of radiation exposure to which it subjects patients.

In the present study we compared preoperative CT and MRI observations with surgical findings.

PATIENTS AND METHODS

In the period 1986-1998, 100 consecutive patients (48 children and 52 adults) received a cochlear implant at our medical center. Nine patients received an extra-cochlear implant (3M House or Med-El) while 91 received an intracochlear device (Nucleus mini-system 22, Laura, Med-El combi 40 or the Advanced Bionics Clarion system) with an electrode array approximately the length of the basal turn of the cochlea.

The mean age at the time of implantation was 26 years; ages ranged from 3 to 68 years. The etiologies of deafness are shown in Table 1. The most common cause of deafness was meningitis, especially in the children (28 of 48).

All the patients underwent high resolution CT scans of both temporal bones with contiguous 1 mm axial sections (Siemens Somatom dr3, Siemens HiQ and Siemens Somatom Plus 4). The examinations were processed on the basis of a bone algorithm. In selected cases, multiplanar reconstructions were made in different planes.

MRI of the inner ear and internal auditory canal has been performed on 28 patients since 1995, using a 1.5 T system (Siemens, Magnetom SP) and a circular head coil. With the aim of detecting intracochlear signal abnormalities, two different high-resolution T2

Cause of deafness	number	surgical class*					
		1	2	3	4	5	other
Meningitis	45	19	3	3	8	9	3
Congenital	9	9	-	-	-	-	-
Usher's syndrome	9	9	-	-	-	-	-
Progressive	6	6	-	-	-	-	-
Hereditary	6	6	-	-	-	-	-
Otosclerosis	4	1	1	1	1	-	-
Enlarged vestibular aqueduct	3	3	-	-	-	-	-
Dysplasia of bony labyrinth	2	2	-	-	-	-	-
Fracture	2	2	-	-	-	-	-
Mumps	2	2	-	-	-	-	-
Ototoxic drugs	1	1	-	-	-	-	-
Waardenburg's syndrome	1	1	-	-	-	-	-
Recurrent ear infections	1	1	-	-	-	-	-
Unknown	9	9	-	-	-	-	-
Total	100	71	4	4	9	9	3

* explanation see text

Table 1 Etiologies of deafness and incidence of surgically encountered compromised cochlear patency.

weighted sequences were used and compared (T2W GE FISP 3D and T2W GE CISS 3D). To assess the neurovascular bundle in the internal meatus, an additional T1 weighed 3D sequence was obtained from the first 22 patients (T1W GE FLASH 3D).

All CT and MRI studies were reviewed by two readers who reached consensus in every case while being kept unaware of the intraoperative findings.

Surgery was performed either by the same experienced otologist or, in some cases, under his supervision. A standard canal wall-up mastoidectomy with posterior tympanotomy was applied to expose the round window niche. The electrodes of the extracochlear implants were placed at that location after identifying the round window membrane. Cochleostomy just anterior to the round window membrane enabled the insertion of the intra-cochlear electrode arrays into the scala tympany. In case of ossification, the scala media and vestibuli were also explored. In totally ossified cochleas, a tunnel was drilled in the direction of the basal turn to enable the insertion of 8-13 electrodes.

The classification according to Parisier[11] was used to describe the amount of ossification encountered intraoperatively: class 1, no ossification present; class 2, round window membrane ossified; class 3, ossification of the round window extending 0-2 mm into the

	CT:					MRI:				
	#	TP	TN	FP	FN	#	TP	TN	FP	FN
Meningitis total:	**45**	**9**	**19**	**0**	**17**	**9**	**4**	**3**	**0**	**2**
class 1:	19	0	19	0	0	3	0	3	0	0
class 2:	3	0	0	0	3	1	0	0	0	1
class 3:	3	0	0	0	3	1	1	0	0	0
class 4:	8	1	0	0	7	2	1	0	0	1
class 5:	9	7	0	0	2	2	2	0	0	0
other:	3	1	0	0	2	0	0	0	0	0
Otosclerosis total:	**4**	**1**	**1**	**0**	**2**	**2**	**1**	**1**	**0**	**0**
class 1:	1	0	1	0	0	1	0	1	0	0
class 2:	1	0	0	0	1	0	0	0	0	0
class 3:	1	0	0	0	1	0	0	0	0	0
class 4:	1	1	0	0	0	1	1	0	0	0
Other etiologies:	**51**	**0**	**51**	**0**	**0**	**17**	**0**	**17**	**0**	**0**
Total:	**100**	**10**	**71**	**0**	**19**	**28**	**5**	**21**	**0**	**2**

#: total number of CT scans or MRI scans, respectively; TP: true positive; TN: true negative; FP: false positive; FN: false negative

Table 2. CT and MRI results related with surgical findings.

scala tympany but coils patent; class 4, ossification of the round window extending 3-8 mm into the scala tympany with patent coils; class 5, diffuse cochlear ossification.

RESULTS

Forty-five patients were deafened by meningitis and 55 by miscellaneous etiologies (Table 1). Reduced cochlear patency as diagnosed intraoperatively was found in 26 of the 45 meningitis patients and in 3 of the 4 patients with otosclerosis. No cochlear obstructions were found with other etiologies. CT was available in all 29 cases with intraoperatively encountered ossification, and in 7 of them MRI as well (Table 2). In the 71 cases in which no ossifications were encountered, a normal patency was diagnosed

	Sensitivity	Specificity	PPV	NPV
CT	34 (78)	100	100 (100)	79
MRI	71 (100)	100	100 (100)	91

In parentheses: same values for the intraoperatively diagnosed class 5 cochleas (meningitis only). Specificity and NPV cannot be calculated for this group. PPV: Positive Predictive Value, NPV: Negative Predictive Value (See also Table 2).

Table 3. Diagnostic value of CT and MRI with regard to cochlear patency of the basal turn in percentages.

correctly by CT (71) and by MRI when available (21) (Specificity of 100%). In 29 cases some degree of ossification was found. As indicated in Tables 1 and 2, CT and MRI findings corresponded poorly with the preoperative assessed patency of the cochlea:

In 4 patients (3 meningitis and 1 otosclerosis), only ossification of the round window membrane was found during surgery (Parisier class 2). This was not detected by CT or MRI. All the electrodes could be inserted after opening the scala tympani.

In 4 patients (3 meningitis and 1 otosclerosis), a class 3 cochlea was found, which again had not been predicted by CT. In this group signs of ossification were seen on the CT scans of 2 of the contralateral cochleas and of the semicircular canals in a third patient. In

Figure 1a. HRCT of both ears. Patient deafened by meningitis. No signs of cochlear ossification visible in the basal turn on the right side.

one of these "class 3" patients an MRI scan had been made which correctly showed a subtle reduction of signal from the basal turn (Figures 1a and 1b). All the electrodes could be inserted after minimal drilling in this group.

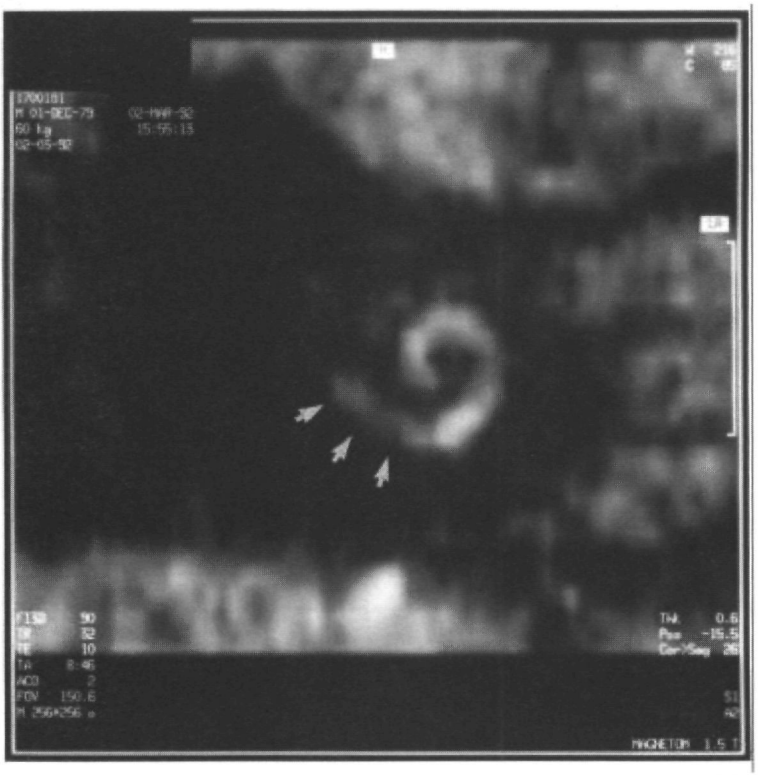

Figure 1b. Same patient. Multiplanar reconstruction parallel to the basal turn of the right cochlea, reconstructed from axial FISP 3D data set. Reduced signal from the beginning of the basal turn (arrows). Per-operatively diagnosed as class 3 ossification, all electrodes could be introduced after minimal drilling.

In only 2 out of the 9 patients (8 meningitis and 1 otosclerosis) with class 4 ossification was obstruction correctly predicted by CT. One of the three available MRI studies also gave a false negative result. In the other 2 cases MRI correctly predicted reduced cochlear patency. CT scans showed signs of ossification in 4 of the contralateral ears in this group. One of the class 4 cases had an etiology of otosclerosis which was clearly demonstrated on the CT scan by the double ring sign but not on the MRI scan although the MRI scan correctly predicted some narrowness of the basal turn (Figures 2a and 2b). Even though in some patients a tunnel of about 6 mm long had to be drilled before open scala was reached, full insertion of the electrodes was possible in all of the patients with class 4 ossification.

Figure 2a. HRCT of both ears after mastoidectomy. Bilateral fenestral and retrofenestral otosclerosis. The double ring sign is demonstrated (arrows). Ill defined basal turn of the cochlea although a lumen remains visible.

Nine patients appeared during surgery to have total obliteration of the cochlea (class 5), which significantly limited the number of electrodes that could be inserted by means of the applied technique (max 13, average 7.3). Total obliteration was correctly predicted by CT in 7 of the 9 patients, although in 2 cases the obliteration did not show. The 2 MRI studies available in this group correctly revealed reduced signal from the basal turn. One of these MRI studies was performed in one of the 2 cases in which the CT was false negative (Figures 3a and 3b). Eight of the nine contralateral inner ears showed ossification on CT and, when available, also on MRI.

In 3 patients, the Parisier classification did not apply ("other" in Table 2). In one of them the CT scan showed ossification of the cochlea which was confirmed during surgery but could not be classified accurately because an extra-cochlear single channel system was placed. In another patient, with a normal CT scan and an intact round window reflex, no ossification was encountered until the apical end of the basal turn (not covered by the classification), which inhibited the insertion of more than 15 electrodes. The third patient also had apical ossification but a normal beginning of the basal turn, which allowed 13

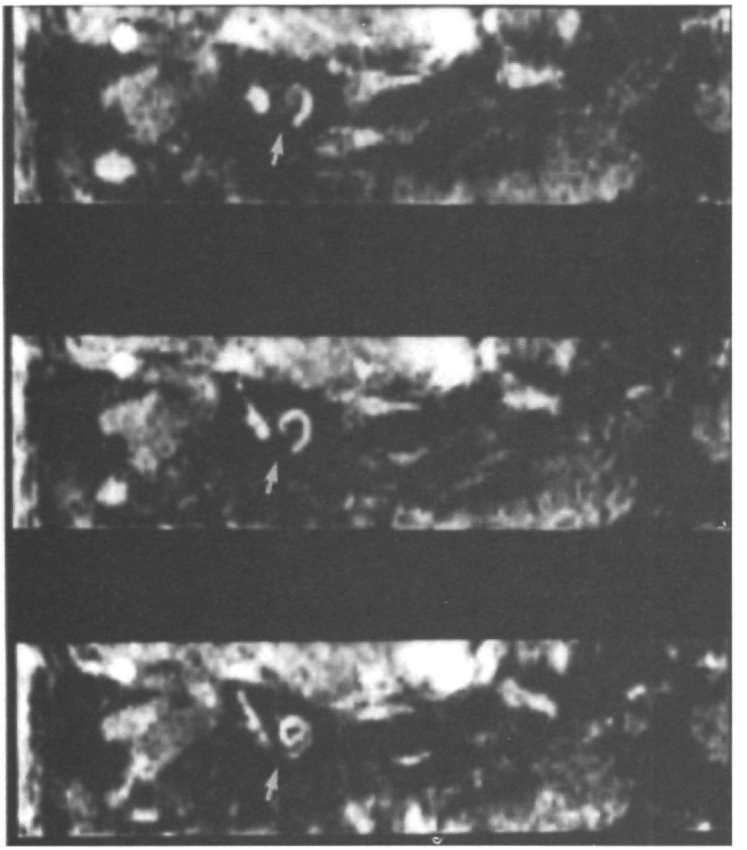

Figure 2b. Same patient. Multiplanar reconstruction parallel to the left basal turn, reconstructed from axial FISP 3D data set. Although MRI does not show specific alterations indicating otosclerosis, focal signal loss of the basal cochlear turn is demonstrated (arrows).

electrodes to be inserted. Apical ossification in the latter patient was not correctly predicted by CT.

DISCUSSION

High-resolution computed tomography provides the cochlear implant surgeon with detailed information about the anatomy of the middle and inner ear structures.[7] It has proven to be a powerful diagnostic tool for all sorts of bony abnormalities, including ossification of the cochlear turns. In the present study of 100 patients, CT proved to be useful for detecting two cases of cochlear dysplasia, three cases of an enlarged vestibular aqueduct, four cases of cochlear otosclerosis, one fracture, some ears with signs of previous surgery and some specific anatomical variations in the position of the sigmoid sinus and

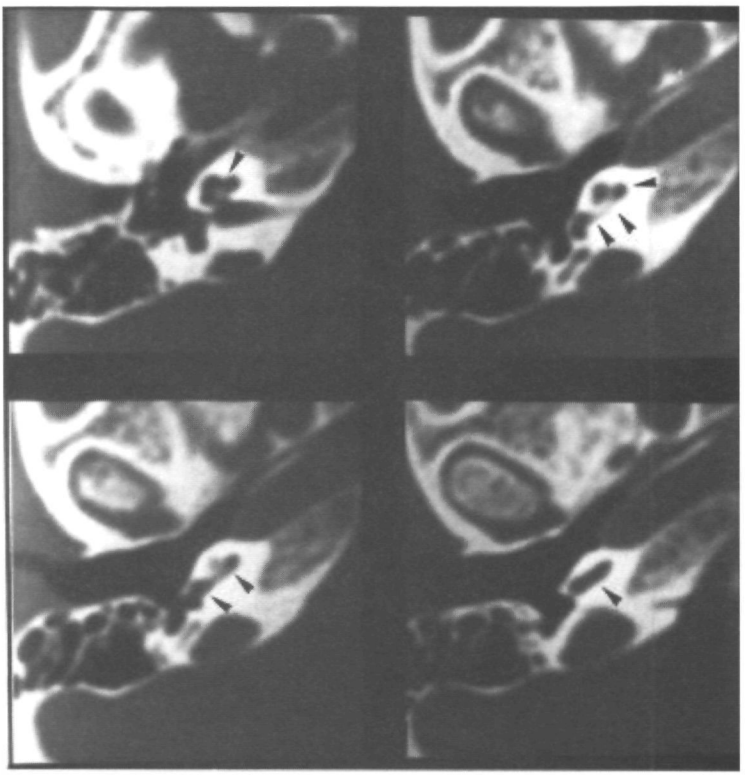

Figure 3a. HRCT of the right ear. Postmeningitis deafness patient. Normal cochlear turns (arrows).

the facial recess area.

None of these etiologies is regarded as a contraindication for cochlear implantation. Partial ossification of the basal turn of the cochlea (classes 2 to 4) is also no longer regarded as a contraindication for placing intracochlear multichannel implants.[2] In most of these cases, multichannel implants resulted in better auditory performance than single-channel systems. However, this is only partially true in patients with totally obliterated cochleas, in whom only a limited number of electrodes could be inserted [4]. Therefore it is important to diagnose severe ossification preoperatively in order to decide whether or not to go through with implantation, to choose the most beneficial implant system, or at the least to be able to bring expectations to a realistic level.

In agreement with previous studies we found that a positive CT scan was a good predictor of compromised cochlear patency (Positive Predictive Value = 100%) but that it gave a relatively large number of false negative results (overall Sensitivity = 38%). This was especially the case with less pronounced ossifications in the basal turn, but also occurred in some cases of total (class 5) obliteration (Sensitivity = 78%). A high percentage of ossified

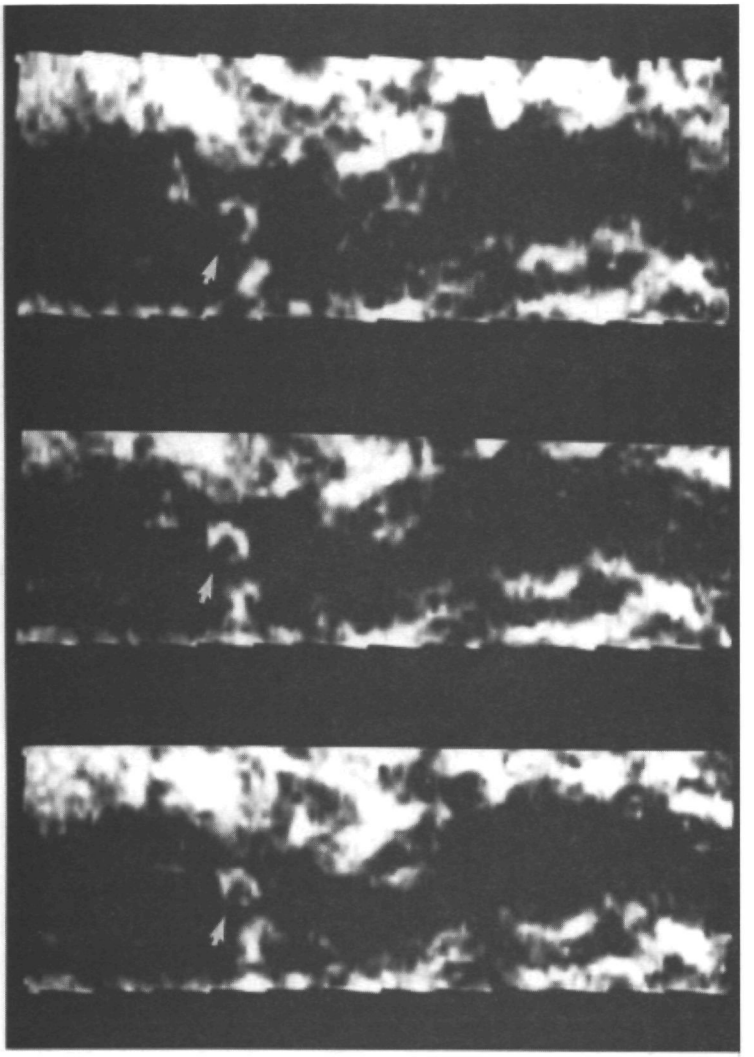

Figure 3b. Same patient. Multiplanar reconstructions parallel to the basal turn of the right cochlea, reconstructed from axial FISP 3D data set. Focal signal loss of the basal turn (arrows). No cochlear lumen could be found during surgery even after extensive drilling (per-operatively classified as class 5 ossification).

ears was seen on the contralateral CT scans of patients with a class 4 or 5 cochlea. Although this is not a reliable predictor of ossification, it can be regarded as a warning sign. Obviously these findings could not be confirmed by surgical exploration. The fact that positive MRI findings are taken into account when choosing the side of implantation also limits the number of MRI studies that can be corroborated with surgical findings. However, not taking this information into account, as in a double blinded study,

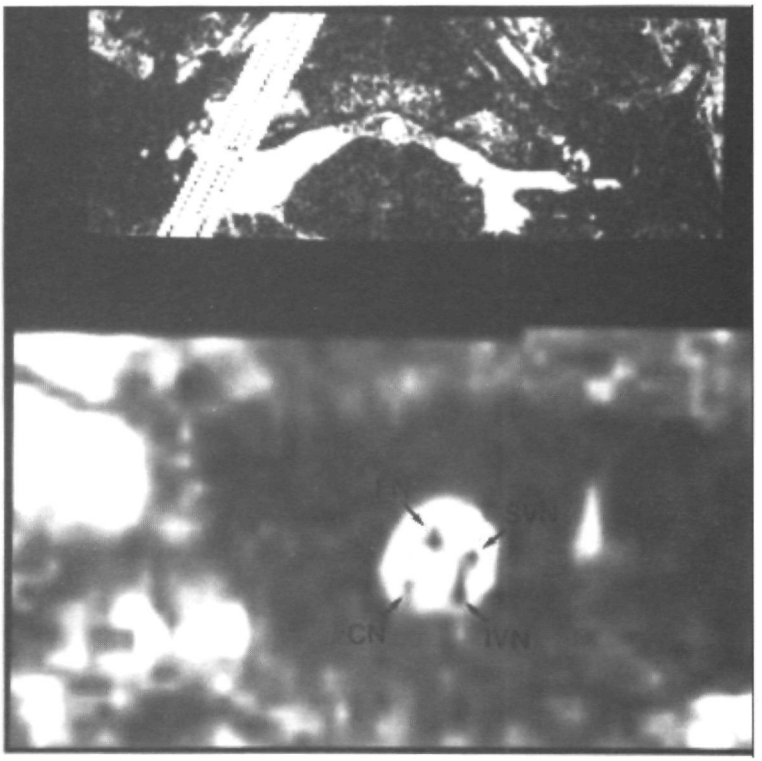

Figure 4. Mutiplanar reconstruction perpendicular to the internal auditory canal (IAC) obtained from axial CISS 3D data set. Good CSF-nerve contrast makes visualisation of the nerves inside the IAC possible. FN=facial nerve, SVN=superior vestibular branch of eighth nerve, CN=cochlear branch of eighth nerve, IVN=inferior vestibular branch of eighth nerve. With the FLASH 3D MRI and the use of multiplanar reconstructions it was possible to distinguish between the seventh and eighth cranial nerves. The branches of the eighth cranial nerve were visible in all cases on the CISS sequence and in most cases on the FISP 3D sequence. Neurinomas or degeneration of the cochlear nerve were not found.

obviously would not have been proper. As this is an inherent shortcoming of all studies dealing with this subject, we felt it to be important to present the available findings in this paper despite the limited number of MRI studies. In contrast to the CT findings, there were no false negative MRI results among the more severe cases of cochlear ossification (Sensitivity = 100%), although MRI failed to predict some cases of ossification restricted to the round window area (overall Sensitivity = 71%). The latter finding is characteristic of MRI which only visualizes fluid-filled cochlear turns instead of the inner ear structures themselves thus obscuring the start of the basal turn. One exception was a class 4 cochlea in which the first 5 mm of the basal turn contained several independent foci of ossification, separated by small parts with a normal lumen, which was not detected by MRI. MRI

seems to be a very reliable predictor of successful implantation, however, because partial electrode insertion was only performed in class 5 cochleas. In this respect it can be argued that the surgical classification for cochlear ossification is of limited clinical value as class 1-4 ossifications pose no serious problems regarding full electrode insertion, considering that it is difficult for the surgeon to differentiate between true ossification and fibrous obstructions. Furthermore, the classification is only applicable for basal cochlear ossifications and omits defining ossifications in the more apical turns.

Congenital abnormalities like cochlear dysplasia and enlarged vestibular aqueducts were visualized equally well by CT and MRI. MRI was not able to detect cochlear otosclerosis or middle-ear and mastoid abnormalities. A considerable advantage of MRI over CT is the accurate visualization of the fluid-filled labyrinthine structures (Figure 4) and its diagnostic value of detecting retrocochlear abnormalities without radiation exposure to the patient. An important advantage of the CISS sequence was the very short aquisition time (only four minutes) which makes this sequence tolerable for most patients and considerably reduces the cost factor.[3]

CONCLUSIONS

Both HRCT and MRI proved to be useful in the preoperative work-up for cochlear implantation. HRCT provided the best information about bony structures, but gave false negative results regarding cochlear patency, even in some cases of more severe obstruction, as was encountered in the meningitis group. MRI was able to detect congenital abnormalities and gave detailed information about the contents of the internal auditory canal and higher retrocochlear structures but did not provide any information about bony structures. MRI appears to be superior to CT with respect to predicting cochlear patency.

In view of the above, starting with MRI rather than CT in the examination of candidates for cochlear implantation who have an etiology of meningitis might be worth considering. Early detection of reduced cochlear patency in the selection procedure for cochlear implantation is important as it can strongly affect the success of the treatment. Obviously, a CT scan can always be performed at a later stage of the selection process. In all other patients, a CT scan is still the diagnostic method of choice to evaluate and detect the presence of anatomical anomalies and otosclerosis.

REFERENCES

1 Arriaga MA, Carrier D MRI and clinical decisions in cochlear implantation Am J Otol 1996,17 547-553

2 Balkany TB, Gantz B, Nadol JB Multichannel cochlear implants in partially ossified cochleas Ann Otol Laryngol 1998,97 3-7

3 Casselman JW Pathology of the membranous labyrinth comparison of T1- and T2-weighed and gadolineum-enhanced fast spin-echo and 3DFT-CISS imaging Am J Neuroradiol 1993,14 59-69

4 Cohen NL, Waltzman SB Partial insertion of the Nucleus multichannel cochlear implant technique and results Am J Otol 1993,14 357-361

5 Frau GN, Luxford WM, Lo WWM, Berliner KI, Telischi FF High-resolution computed tomography in evaluation of cochlear patency in implant candidates a comparison with surgical findings J Laryngol Otol 1994,108 734 748

6 Green JD, Marion MS, Hinojosa R Labyrinthitis ossificans histopathologic considerations for cochlear implantation Otolaryngol Head Neck Surg 1990,104 320 326

7 Jackler RK, Dillon WP Computed tomography and magnetic resonance imaging of the inner ear Otolaryngol Head Neck Surg 1998,99 494-504

8 Johnson MH, Hasenstab MS, Seicshnaydre MA, Williams GH CT of postmeningitic deafness observations and predictive value for cochlear implants in children Am J Neuroradiol 1995,16 103-109

9 Klein HM, Bohndorf K, Hermes H, Schutz WF, Gunther RW, Schondorff G Computed tomography and magnetic resonance imaging in the preoperative work-up for cochlear implantation Eur J Rad 1992,15 89-92

10 Masot-Dupuch K, Chouard CH, Falisse B, Meyer B, Tubiana JM Place of 3DFT-MR imaging study on cochlear implant candidates In Fraysse B, Deguine O (eds) Cochlear implants new perspectives 1993 Karger, Basel, p 23-28

11 Parisier SC, Chute PM Multichannel implants in postmeningitic ossified cochleas In Fraysse B, Deguine O (eds) Cochlear implants new perspectives 1993 Karger, Basel, p 49-58

12 Seicshnaydre MA, Johnson MH, Hasenstab MS, Williams GH Cochlear implants in children reliability of computed tomography Otolaryngol Head Neck Surg 1992,107 410-417

13 Seidman DA, Chute PM, Parisier S Temporal bone imaging for cochlear implantation Laryngoscope 1994,104 562-565

14 Silberman B, Garabedian EN, Denoyelle F, Moatti L, Roger G Role of modern imaging technology in the implementation of pediatric cochlear implants Ann Otol Rhinol Laryngol 1995,104 42-46

15 Wiet RJ, Pyle GM, O'Connor CA, Russell E, Schramm DR Computed tomography how accurate a predictor for cochlear implantation? Laryngoscope 1990,100 687-92

CHAPTER THREE

THE RISK OF VESTIBULAR FUNCTION LOSS AFTER INTRACOCHLEAR IMPLANTATION

P. L. M. Huygen, J. B. Hinderink, P. Van Den Broek, S. Van Den Borne, J. P. L. Brokx, L. H. M. Mens and R. J. C. Admiraal

Acta Otolaryngology (Stockholm) 1995; Suppl 520: 270-272

THE RISK OF VESTIBULAR FUNCTION LOSS AFTER INTRACOCHLEAR IMPLANTATION

P. L. M. Huygen, J. B. Hinderink, P. Van Den Broek, S. Van Den Borne, J. P. L. Brokx, L. H. M. Mens and R. J. C. Admiraal

ABSTRACT

Sixty patients were selected for cochlear implantation and 50 of them received an intracochlear implant (Nucleus) Vestibular function was evaluated before and after surgery using a caloric test and a velocity step test Sixteen patients had normal or residual vestibular function before surgery, 11 bilateral and 5 unilateral, in 3 of the latter patients, the ear with vestibular areflexia was elected for implantation, which reduced the number of patients at risk for vestibular dysfunction to 13 Vestibular function was preserved in all of these patients except for 4, the risk of vestibular function loss can therefore be rated at about 31% Key words deafness (acquired, genetic), vestibular areflexia, vestibular hyporeflexia

INTRODUCTION

Only a few reports have appeared on the results of vestibular tests in relation to intracochlear implantation.[1-5] According to our previous reports[6,7] on our own (preliminary) data and other reported data, the risk of vestibular function loss can be estimated at between 50 and 60%. Since the submission of our previous reports, several new patients have been implanted at our department and our current data indicate that the risk of vestibular function loss may be lower.

MATERIALS AND METHODS

Our study population comprised 60 patients, 50 of whom received an intracochlear implant, i.e. a Nucleus device (Cochlear Corporation, Englewood, Colorado, USA). Extracochlear implantation was performed in the early period (1987-1990) and, more recently, in a patient with Mondini-type dysplasia. The patients (30 males, 30 females) were aged between 5 and 68 years; 18 of them were younger than 13 years. The aetiological diagnoses of these patients are presented in Table 1. The methods used to evaluate vestibular function and the classification into the categories vestibular areflexia, hyporeflexia, normoreflexia (and hyperreflexia) have been described previously).[6,7]

RESULTS

Preimplant findings (60 patients)

None of our patients showed any gaze-evoked nystagmus or spontaneous nystagmus. Smooth pursuit and optokinetic nystagmus (OKN) responses were normal in 58 of the patients. In the remaining 2 who had Usher's type I syndrome, the OKN response levels were too low (they had constricted visual fields and poor visual acuity). Table 1 shows the preimplant findings in our patients according to their aetiology (in 1 child after meningitis it could not be concluded whether normo- or hyperreflexia applied to the sinusoidal responses and caloric tests were omitted). Vestibular areflexia manifested itself in 38 patients (63%) as a total lack of nystagmus after velocity step tests of 90"/s (plus 250"/s in 3 cases).

Findings after intracochlear implantation (50 patients)

Of the 22 patients with preimplant vestibular (hypo-or normal) function, 2 received extracochlear implants, 4 were not evaluable (3 had severe hyporeflexia and caloric tests were therefore omitted and 1 had an abnormally shaped semicircular canal which was inadvertently opened during surgery causing vestibular loss). All of the 16 remaining

patients underwent intracochlear implantation and their preimplant vestibular function could be evaluated (Table 2). Three of these patients were not at risk because the ear

Aetiology	Vestibular function			
	Bilateral areflexia	Hyporeflexia	Normoreflexia	Hyperreflexia
Meningitis	27	2		
Mumps		1		
Head trauma	2			
Ototoxicity				2
Unknown		4	3	
Congenital severe SNHL				
Usher I (276900)	6			
Mondini dysplasia		1		
AD (124580)			1	
AR(220700, 800)	2			
Progressive SNHL				
Otosclerosis	1	1	1	
AD hf (124800)			2	
AR (221650)				1
AR? unidentified		1	1	
Total	38	10	8	3

Table 1. Preimplant findings by aetiology
AD=autosomal dominant; AR=autosomal recessive; hf=high frequency; SNHL=sensorineural hearing loss (MIM number)[10]

elected for implantation had complete vestibular function loss.

Table 3 shows that 11 of the 13 remaining patients whose vestibular function was at risk on the side of implantation, were at risk of developing unilateral loss (their preimplant function had been intact bilaterally). Three of these patients had a vestibular deficit following implant surgery. One of them did not experience any appreciable symptoms, presumably because the lost labyrinth had already shown reduced sensitivity before implantation (the caloric response level was 56% of that obtained from the other labyrinth). The other 2 patients had the classical symptoms of a unilateral vestibular deficit. Eight patients had a repeat vestibular examination which showed complete preservation of vestibular function in the implanted ear. One of them had vestibular complaints and showed hyperactive velocity step responses postimplant, but she had displayed similar findings before implantation, which could be attributed to hyperventilation;[8] physical breathing control therapy was recommended

In 2 patients with unilateral function loss, the other labyrinth was at risk because it had been elected for implantation. After the implantation, one of them revealed bilateral vestibular areflexia with the associated typical symptoms[9] and the other had intact function.

A total of 13 patients who were at risk of losing vestibular function in the ear elected for implantation could be evaluated. Four out of these 13 patients lost their function. Therefore, the risk of losing vestibular function through intracochlear implantation can be rated at 4 out of 13, or about 31%.

Initial number of patients	22
Extracochlear	-2
Not evaluable	-4
	16
Evaluable	
Not at risk of losing bilateral	-3
Evaluable at risk	13

Table 2. Patients with normal or residual vestibular function preimplant

DISCUSSION

During intracochlear implantation, the electrode is inserted through the round window and led into the scala tympani over a length of some 2 cm. This procedure may damage the basilar membrane or the spiral ligament and this carries the risk of endolymph mixing with perilymph with subsequent loss of inner ear functions. At present, our results indicate a risk of about 31%, which is somewhat lower than the 50-60% mentioned in our previous reports. [6,7] Nevertheless, we are of the opinion that the patient should be informed beforehand about this risk – if applicable – and the possible consequences of vestibular areflexia.[9] The same applies to the impending risk of unilateral function loss, although it seems reasonable to suppose that this would mean a much less severe handicap to most patients. In one of our patients, the preimplant caloric sensitivity on the side that was later elected for implantation was hardly more than half of that on the other side; the total unilateral loss of vestibular function which occurred after implantation took a subclinical course and the patient remained asymptomatic.

The present selection of cochlear implant candidates offers some indication as to what can be expected to happen to vestibular function in relation to aetiology in similar cases. On the one hand, bilateral vestibular areflexia, by definition, is to be found in Usher's type I syndrome[10] ("Mendelian inheritance in Man" or MIM number 276900[11]) and it generally occurs in patients with bilateral deafness following meningitis[12] or head trauma. On the other hand, the autosomal dominant (AD)

Exposed at risk	Function lost	Function intact
At risk of losing unilateral function: 11	3[a]	8
At risk of losing bilateral function: 2	1[b]	1
Total: 13	4 (31%)	9

[a]1 symptom-free (preimplant function already partially lost).
[b]All the symptoms of total vestibular areflexia.[9]

Table 3. Outcome in at-risk patients

syndrome of progressive sensorineural hearing loss (SNHL) which starts in (early) childhood at the high frequencies (MIM 124800) is generally associated with normal vestibular function[12, 13 and additional unpublished data] and, presumably, this also applies to autosomal recessive (AR) progressive SNHL with childhood onset (MIM 221650).[14 and additional unpublished data] In other categories of patients, e.g. with acquired bilateral SNHL, congenital AD SNHL (MIM 124580), congenital AR severe SNHL (MIM 220700, 220800[12] and otosclerosis[11]), it is uncertain what will happen to their vestibular function. We are therefore of the opinion that vestibular examination should be performed as an integral part of the selection procedure of all prospective candidates for implantation, because at the very least it will help to avoid the development of bilateral areflexia in some patients.

REFERENCES

1 Black FO Present vestibular status of subjects implanted with auditory prostheses Ann Otol Rhinol Laryngol 1977,Suppl 38 49-56
2 Chouard CH, Fugain C, Meyer B, Lacombe H Resultats cliniques de l'implant cochleaire a multi electrodes Ann Otolaryngol (Paris) 1981,98 593-612
3 Eisenberg LS, Nelson JR, House WF Effects of the single-electrode cochlear implant on the vestibular system of the profoundly deaf adult Ann Otol Rhinol Laryngol 1982,Suppl 91 47-54
4 Black FO, Lilly DJ, Peterka RJ, Fowler LP, Simmons FB Vestibulo-ocular and vestibulospinal function before and after cochlear implant surgery Ann Otol Rhinol Laryngol 1987,Suppl 128 106-9
5 Mangham CA Effects of cochlear prostheses on vestibulo-ocular reflexes to rotation Ann Otol Rhinol Laryngol 1987,Suppl 128 101-4
6 Van den Broek P, Huygen PLM, Mens LHM, Admiraal RJC, Spies T Vestibular function in cochlear implant patients Acta Otolaryngol (Stockh) 1993,113 263-5
7 Huygen PLM, van den Broek P, Spies TH, Mens LHM, Admiraal RJC Does intracochlear implantation jeopardize vestibular function? Ann Otol Rhinol Laryngol 1994,103 609-14
8 Theunissen EJJM, Huygen PLM, Folgering HT Vestibular hyperreactivity and hyperventilation Clin Otolaryngol 1986,11 161-9
9 Gresty MA, Hess K, Leech J Disorders of the vestibulo-ocular reflex producing oscillopsia and mechanisms compensating for loss of labyrinthine function Brain 1977,100 693-716
10 McKusick VA Mendelian inheritance in man Catalogs of autosomal dominant, autosomal recessive, and X-linked phenotypes, 10th ed Baltimore Johns Hopkins University Press, 1992
11 Huygen PLM, Verhagen WIM Peripheral vestibular and vestibulo-cochlear dysfunction in hereditary disorders A review of the literature and a report on some additional findings J Vestib Res 1994,4 81-104
12 Huygen PLM, Van Rijn PM, Cremers CWRJ, Theunissen EJJM The vestibulo-ocular reflex in pupils at a Dutch school for the hearing impaired, findings relating to acquired causes Int J Pediatr Otorhinolaryngol 1993,25 39-47
13 Gorlin RJ, Toriello HV Hereditary hearing loss and its syndromes New York Oxford University Press, 1994
14 Cremers CWRJ Autosomal recessive nonsyndromal progressive deafness in childhood A separate clinical and genetic entity Int J Pediatr Otorhinolaryngol 1979,1 193-9

CHAPTER FOUR

RESULTS FROM FOUR COCHLEAR IMPLANT PATIENTS WITH USHER'S SYNDROME

Johannes B. Hinderink, MD, Jan P.L. Brokx, PhD,
Lucas H.M. Mens, PhD, Paul van den Broek, MD, PhD

Annals of Otology, Rhinology and Laryngology 103: 1997 pp. 285-293

RESULTS FROM FOUR COCHLEAR IMPLANT PATIENTS WITH USHER'S SYNDROME

Johannes B. Hinderink, MD, Jan P.L. Brokx, PhD,
Lucas H.M. Mens, PhD, Paul van den Broek, MD, PhD

ABSTRACT

Individual results are presented of four patients with Usher syndrome type I who received a cochlear implant Both single-channel and multichannel implants were used Because of implant failure, one of the single-channel systems was replaced by a Nucleus multichannel system Results are compared to the results of five other prelingually deaf cochlear implant users The performance of the patients with Usher's syndrome on suprasegmental and segmental speech perception tests and on a connected discourse tracking task did not differ significantly from the performance of the other prelingually deaf patients A significant improvement over time was found at the suprasegmental level for the combined group of Usher's and other patients No obvious differences were found between the scores from the patients with a single-channel and the patients with a multichannel device The rehabilitation of the Usher's patients required very little extra effort in comparison with that of the other prelingually deaf patients, all patients reported considerable advantages in hearing abilities and social life

INTRODUCTION

In the past decade, a large number of studies have documented that cochlear implantation improved the auditory perception of the majority of adult postlingually deaf patients.[1-3] Although, according to these studies, the speech perception results of prelingually deaf adult implant users are generally much poorer than those of postlingually deaf adult users, various tests demonstrated some improvement of speech perception, and many authors reported subjective benefits for most of the prelingually deaf patients.[4-7] Nevertheless, it is not generally agreed that these benefits justify implanting a large number of prelingually deaf adults.

In prelingually deaf patients with multiple sensory deficits, for example patients with Usher's syndrome who are deaf and also suffer from defective vision, it would be worthwile to make an inventory of the objective and subjective benefits of cochlear implantation, because it is assumed that the signal function of the cochlear implant can provide great practical and psychologic support for these patients.

The prevalence of Usher's syndrome in congenitally deaf patients is reported to range from 3% to 6% and it is the leading cause of combined deafness and blindness in the United States.[8] Only a few studies explicitly describe small numbers of implanted patients with Usher's syndrome,[6,9] and only Dawson et al[6] used objective speech perception tests. However, no comparison was made between the results obtained with a single-channel implant and those obtained with a multichannel system, and no details were given about the occurrence of specific rehabilitation problems in implanted Usher's patients. Eisenberg[9] reported on 2 patients with Usher's syndrome who were using single-channel implants. Despite their background in manual communication, the patients described by Eisenberg were using the implant on a daily basis. Furthermore, subjective benefits of the implant were mentioned, such as being able to respond to attention-getting sounds, improvement of their own voice quality, and feeling more involved socially and less dependent on others. No objective data were presented on speech perception abilities. Dawson et al[6] described the speech perception performance of 3 patients with Usher's syndrome (1 adolescent and 2 adults) who were using the Nucleus multichannel implant and compared their performance to that of young prelingually deaf implanted children. In contrast to the children's performance the older patients did not demonstrate any open-set speech recognition or improvement over 2 to 3 years of postoperative follow-up.

These studies, however, do demonstrate that the results of the patients with Usher's syndrome are similar to those of other prelingually deaf patients. This is in agreement with the results of case studies on blind cochlear implant users who showed significant improvement in speech perception.[10] It should be noted that despite severe visual impairment, most of the patients with Usher's syndrome who have been included in implant trials still had enough functional central vision to be able to lip-read, read and

Patient	Sex	Cause of Deafness	Onset of Deafness	Preoperative PTA	Age at implantation	System used
A	F	Usher	congenital	>120	20y1mo	1
					23y2mo*	22*
B	M	Usher	congenital	>110	13y5mo	22
C	M	Usher	congenital	>120	28y11mo	1
D	F	Usher	congenital	>120	20y7mo	22
E	M	meningitis	3 mo.	>120	20y6mo	1
F	F	meningitis	3 mo.	>120	10y1mo	1
G	M	meningitis	2 y 3 mo.	>120	19y6mo	1
H	F	Mondini	congenital	>120	24y3mo	1
I	M	hereditary	congenital	>120	33y5mo	22

PTA: Pure Tone Average at 500, 1000 and 2000 Hz in decibels hearing level, 1= single-channel system, 22=multichannel system.
* Reimplantation

Table 1. Patient Data

write.

We performed a speech perception study on 4 patients with Usher's syndrome who were implanted and rehabilitated by a team from the University Hospital Nijmegen and the Institute for the Deaf in St Michielsgestel, the Netherlands. Their speech perception performance after implantation was compared to the results of four other prelingually deaf implant users with different disorders. One of the 4 patients was reimplanted with the Nucleus Mini System 22 after her initial single-channel extracochlear system failed.

PATIENTS AND METHODS

Patients

Some patient characteristics are given in Table 1. The mean age of the Usher's patients and the other prelingually deaf patients at the time of implantation was 21.2 years and 21.9 years, respectively. The length of follow-up was 2 years, except for patient I, who was followed up for 1 year. All the patients were (former) pupils of the Institute for the Deaf and had been enrolled in oral-aural communication programs. The patients with Usher's syndrome (patients A to D in Table 1) are discussed individually below.

Selection

A full description of the selection procedure is given elsewhere.[11] The same selection criteria were applied to the Usher's patients and the other prelingually deaf patients. They all had to be trained lip-readers who were eager to use oral-aural communication. Electrical stimulation by means of a needle electrode placed onto the round window or an electrode that had been placed temporarily in the round window niche[12] had to produce hearing sensations. The availability of a close friend or relative to help them with the rehabilitation

program was required. Any patients with other mental or sensory deficits besides the visual impairment due to Usher's syndrome were excluded. The 4 patients with Usher's syndrome all lacked vestibular function.

Implantation

The single-channel systems (Med-El E/1 or 3M/Vienna) were implanted extracochlearly in the round window niche, with the reference electrode inserted into the bone superior to the lateral semicircular canal. Of the Nucleus systems, all the electrodes could be fully inserted into the cochlea.

Rehabilitation

After a recovery period of at least 4 weeks the processors were fitted. The Nucleus processors were fitted in the bipolar plus 1 mode with all the electrodes active. The subjects and their assistants were then admitted to the Institute for the Deaf for a period of two weeks to take part in the initial phase of an intensive rehabilitation program. Every two months during the first year of follow-up, the patients underwent a 1-day training and testing session and occasional processor readjustment. Rehabilitation of the adolescent patients (B and F) was largely carried out by their own school teachers and was integrated into the regular educational programs at the Institute for the Deaf.

Auditory tests

The auditory and speech perception tests were administered to the patients preoperatively and at 3, 6, 12 and at 24 months after the initial fitting of the processor.

Preoperatively, pure tone audiometry and speech audiometry were performed with standard equipment and procedures. Free field audiometry was applied to obtain thresholds with hearing aids and cochlear implants.

The auditory perception of segmental and suprasegmental aspects of speech was assessed by means of a Dutch version of the Monosyllable-Trochee-Spondee (MTS) test[13] and by means of the Antwerp-Nijmegen (AN) test battery.[14,15] The AN test battery has a set up similar to the MAC and Iowa test batteries.[16,17] It comprises tests for speech recognition (ie, Short Vowel Identification, Long Vowel Identification, Monosyllabic Word Identification, and Spondee Identification), pattern recognition (ie, Number of Syllables, Sentence Accent Identification, Male/Female/Child Discrimination), and an Environmental Sounds Identification test. All the AN tests were in a closed-set format.

Lip-reading skills were tested with a Connected Discourse Tracking (CDT) task[18] in a visual-only condition and an auditory-plus-visual condition. If possible, the CDT task was also performed in an auditory-only condition. A preoperative assessment was made of the number of words per minute that each patient could read aloud from a printed text, ie, the "top score".

Preoperatively and at the 1-year follow-up, an assessment was made of the subjective

aspects of implantation by means of the "Gestel-Nijmegen implant questionnaire". This questionnaire examined hearing abilities and social functioning before and after implantation.

Patient	interval	Frequency in Hz 250	500	1,000	2,000	4,000	8,000
A	Preoperatively	>110	>120	>120	>120	>110	>100
	Hearing aid	65	65	>120	>120	>100	>100
	1-Channel implant	40	65	60	60	75	85
	22-channel implant	40	40	35	35	35	35
B	Preoperatively	80	100	110	>120	>110	>100
	Hearing aid	50	65	65	80	>100	>100
	22-Channel implant	50	65	50	55	50	60
C	Preoperatively	95	120	>120	>120	>110	>100
	Hearing aid	NA	NA	NA	NA	NA	NA
	1-Channel implant	40	45	50	60	45	35
D	Preoperatively	>110	>120	>120	>120	>110	>100
	Hearing aid	75	110	>110	>110	>100	>100
	22-Channel implant	45	40	40	30	30	30
E	Preoperatively	105	115	120	>120	>110	>100
	Hearing aid	NA	NA	NA	NA	NA	NA
	1-Channel implant	50	55	55	55	60	60
F	Preoperatively	115	>120	>120	>120	>110	>100
	Hearing aid	NA	NA	NA	NA	NA	NA
	1-Channel implant	55	65	65	60	70	80
G	Preoperatively	95	120	>120	>120	>110	>100
	Hearing aid	NA	NA	NA	NA	NA	NA
	1-Channel impant	40	45	50	50	45	35
H	Preoperatively	110	120	>120	>120	>110	>100
	Hearing aid	55	60	60	95	>100	>100
	1-Channel implant	35	40	40	35	40	35
I	Preoperatively	105	>120	>120	>120	>110	>100
	Hearing aid	70	60	80	>110	>100	>100
	22-Channel implant	40	45	40	30	40	45

Preoperative unaided thresholds of best ear and free field aided thresholds (hearing aid or implant) are given in decibels hearing level Patients A-D=Usher syndrome, patients E-I=other prelinguals Letters NA indicate that threshold was not measured at this frequency Symbol > indicates that output limit of the audiometer was reached without finding a threshold One-year postoperative thresholds with implants are given, except in patients H and I, for whom 6 month thresholds are given

Table 2. Hearing thresholds

Materials

Audiometry was performed in a special sound-proofroom with a standard audiometer (Interacoustics AC-5) calibrated according to ISO 389. For free field audiometry a loudspeaker was placed 1 m in front of the patient. The free field set-up for warble tones was calibrated according to Morgan et al.[19] A tape recorder (Sansui D-35BF), coupled to the audiometer, was used to present the recorded AN tests in the free field setup described

above at a loudness level which was subjectively found to be comfortable.

In the MTS and the CDT tests live speech of one of the rehabilitation therapists was used in a quiet, well-lit room at approximately 1 m from the patient, with a normal conversation level of approximately 65 dB sound pressure level (SPL). During all the tests, the sensitivity of the speech processor was fixed at the normal setting for daily use.

Data analysis

Analysis of variance (General Linear Models procedure, SAS Institute) was used to evaluate the different test scores. Independent factors were 1) cause of deafness (Usher's syndrome versus other causes) and 2) test interval (preoperatively and at 3, 6, 12 and 24 months). The patients formed random variables, nested in Cause of deafness. For the analysis of the AN tests, the type of subtest was included as an independent factor. Significant main effects or interactions were analysed post-hoc with the Student-Newman-Keuls multiple range test (alpha=5%; procedure General Linear Models-Student-Newman-Keuls, SAS Institute).

RESULTS

Case Report of Patients with Usher's Syndrome

Patient A was born in 1968. She was a pupil at the Institute for the Deaf from 1972 till 1986. She had had profound hearing loss since birth (pure tone average [PTA] over 120 dB hearing level [HL]). She had used a body-worn hearing aid in the right ear till 1981. Retinitis pigmentosa brought about progressive peripheral vision loss that stabilized in 1985 and left her with only 10° of central vision. Promontory stimulation of the right ear in 1986 produced no hearing sensations. In 1988 the temporary insertion of an electrode in the left ear successfully achieved hearing sensations and showed a satisfactory dynamic range (11 dB at 63 Hz) at the end of a 3-week testing period. In 1988, at the age of 20 years, she received an extracochlear single-channel system (3M/Vienna) in the left ear. After almost 3 years of use, her perception abilities were reduced, and finally no signal at all could be perceived. Surgical inspection under local anaesthesia showed that the electrodes were in place and undamaged. It was decided to replace the device with a Nucleus multichannel implant in the same ear. She had been using this implant successfully for 18 months at the time of the last evaluative tests.

In Table 2 the hearing loss, the preoperative aided thresholds with the previous hearing aid, and the 1-year postoperative thresholds with the single-channel and the multichannel implants are presented. With either implant, the thresholds were markedly improved over the unaided thresholds. At the higher frequencies, considerably better thresholds were achieved with the Nucleus device than with the single-channel implant.

The Dutch version of the MTS Test became available one year after this patient's first implant. In the first testing session she scored significantly above chance on both the syllable

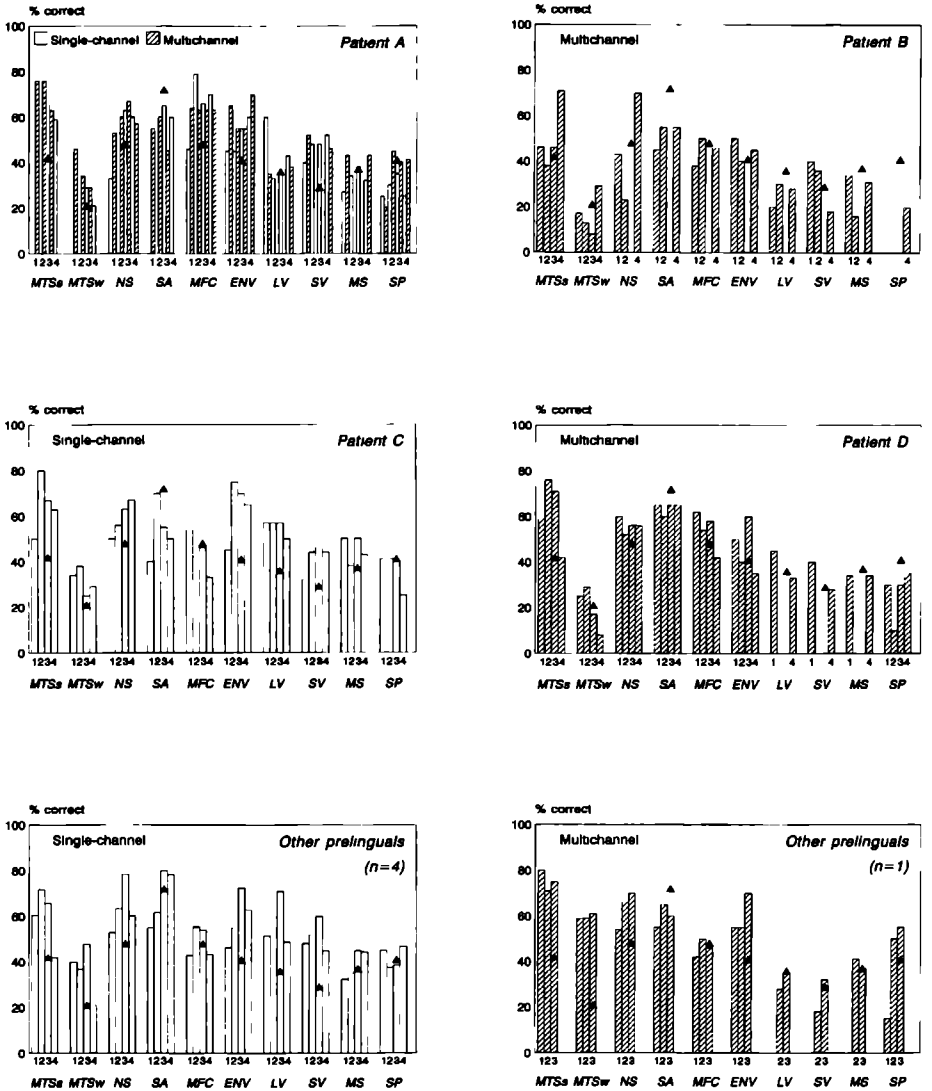

Figure 1. Results on Monosyllable-Trochee-Spondee tests and on tests of Antwerp-Nijmegen battery at 3, 6, 12, and 24 months (1, 2, 3, 4) after implantation. MTSs – MTS syllable detection, MTSw – MTS word recognition, NS – Number of Syllables Identification, SA – Sentence Accent Identification, MFC – Male/Female/Child Discrimination, ENV – Environmental Sounds Identification, LV – Long Vowel Identification, SV – Short Vowel Identification, MS – Monosyllabic Word Identification, SP – Spondee Identification, black triangles – 95% confidence levels. For patient A, scores with both single-channel and multichannel implants are shown.

detection level and the word recognition level (Fig 1) After two years of implant use, the scores were slightly lower Her initial performance on this test with the Nucleus implant was better than that with the single-channel implant However, after 1 year of using the Nucleus implant, the result was comparable to that obtained with the single-channel system

On the AN battery she scored significantly above chance on the following test the Number of Syllables Identification, the Male/Female/Child Discrimination, and the Environmental Sounds Identification tests (Fig 1) Only the scores on the Sentence Accent Identification test were not significantly above chance On the speech recognition tests of the AN battery, her scores with the single-channel implant were significantly above chance, except for the Monosyllabic Word Identification test With the Nucleus implant, her scores were significantly above chance for all 4 tests In sum, the performance with the single-channel amd the multichannel implants were comparable

Scores in the auditory-plus-visual condition of the CDT Task with the single-channel implant approached the "top score," which was 68 words per minute Performance in the audiory-plus-visual condition was slightly better than in the visual-only condition in all the testing sessions This was also the case when the Nucleus implant was used, but the differences between the auditory-plus-visual and the visual-only conditions were smaller (Fig 2)

One year after implantation, patient A was using the single-channel implant on a daily basis in all situations (from 3 to 12 hours a day). She reported that being able to hear and recognize a large variety of environmental sounds made her feel less isolated She also felt more secure and less tense when negotiating traffic. She found listening to music especially pleasant During communication with other people (both hearing-impaired and with normal hearing) she felt more relaxed and could make new contacts more easily The same benefits were reported after one year of using the Nucleus implant, but music received less emphasis Overall she felt more self-confident and more secure with either type of implant and found that the implant had added a great deal of quality to her life

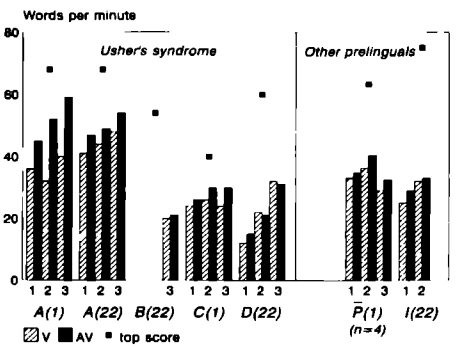

Figure 2 Results on Connected Discourse Tracking task for patients with Usher's syndrome (A to D) and for other prelingually deafened patients (P and I) 1 – Single-channel systems, 22 – multichannel systems, V – visual only, AV – auditory plus-visual, 1, 2, 3 – 3, 6, and 12 months after implantation, respectively

Patient B(PTA over 110 dB HL), was born deaf in 1976. He was a pupil at the Institute for the Deaf from 1984 to 1992 after 4 years in a total communication program. He had been using two behind-the-ear hearing aids up to implantation Deterioration of peripheral

vision stabilized in 1988 and left him with approximately 10° of central vision. Owing to his young age no conventional promontory stimulation was performed. Instead an electrode was placed temporarily in his left ear. This yielded hearing sensations and perception of environmental sounds. Over a 3-week period, the dynamic range increased considerably, from 5 to 21 dB at 63 Hz. In January 1990 at the age of 13.5 years he was implanted with a Nucleus multichannel implant in his left ear. Speech processor programing was difficult because of inconsistent responses and the wish to avoid loud sensations. He had been using the implant for more than 1 year before it appeared that the stimulus current level of one of the electrodes was set too high, causing him to lower the sensitivity setting of the processor after each adjustment. After this was corrected, satisfactory programming was achieved.

Preoperative and postoperative (after satisfactory programming was achieved) hearing thresholds are shown in Table 2. Better thresholds were obtained with the implant than with a hearing aid, especially at the higher frequencies.

At the 2-year testing session, the patient scored significantly above chance on the word recognition level of the MTS test. Although significant scores on the syllable detection level were found in the 1-year testing session, the largest improvement was seen in the second year (Fig 1).

No significant scores were obtained on the Sentence Accent Identification tests of the AN battery (Fig 1), but significant performance was demonstrated on the Number of Syllables Identification, the Male/Female/Child Discrimination and Environmental Sounds Identification tests. On the speech recognition level, only his Short Vowel Identification test performance in the first two testing sessions was significantly above chance.

The CDT task was not part of the protocol for adolescents, so it was not administered to patient B until he had been using the implant for 2 years. No significant benefit of the implant could be demonstrated on this test (Fig 2).

After 2 years of using the implant, patient B and his parents reported many advantages of the implant over the previously used hearing aids. Although the initial processor adjustments were not optimal, many environmental sounds could be detected and recognized with the implant, and communication with other people became easier. Patient B was using his implant all day; he felt safer and more independent. One and a half years after the implant, he successfully enrolled at a school for children with normal hearing but sometimes still had to rely on the help of an interpreter.

Patient C was born deaf in 1961 (PTA over 120 dB HL). He had been using bilateral hearing aids since 1963. He had only 5° of central vision when he entered the selection procedure. Electrical round window stimulation resulted in indistinct sensations and a very small dynamic range. Temporary placement of an electrode in the right ear produced hearing sensations and an increasing dynamic range over time. In 1990, at the age of 29 years he was implanted with a single-channel extracochlear Med-El E/1 system in his right ear. During the first year after the implantation, his peripheral vision diminished even further, which caused noticeable problems during the follow-up rehabilitation sessions.

Preoperative audiometry showed no residual hearing (Table 2). One year after the implantation, his average hearing threshold at 0.5, 1 and 2 kHz was 52 dB HL.

Before implantation, it was impossible for him to perform the MTS test because of profound deafness. In all the postoperative testing sessions, his MTS test scores were significantly above chance at both the syllable and word recognition levels (Fig 1).

Significant scores were obtained on all the tests of the AN battery, with the exception of the Sentence Accent Identification test. Significant scores were obtained for the Spondee Identification test, except during the 12 and 24 month testing sessions (Fig 1).

A poor top score of 40 words per minute was achieved on the CDT task, which reflected his visual restrictions while reading text. In the visual-only condition an average performance of 25 words per minute was measured compared to 27 words per minute in the auditory-plus-visual condition (Fig 2).

Follow-up rehabilitation was troubled by further deterioration of the visual handicap, and this patient therefore expressed the need for a prolonged rehabilitation period. He could hear and recognize a number of environmental sounds and found music very enjoyable. Communication with deaf people and with people with normal hearing was reported to have improved but was still very difficult. He often had to rely on writing or the help of a familiar person when communicating with unknown people. Although he was disappointed about the communication abilities with the implant, he was using the implant in all situations and felt more secure and less isolated with it.

Patient D was born in 1970. She had been using a body-worn hearing aid since profound deafness was diagnosed in 1973 (PTA over 120 dB HL). In 1977, Usher's syndrome was diagnosed. The progressive loss of peripheral vision stabilized during later years, leaving a visual field of approximately 45°. Round window stimulation successfully achieved hearing sensations with a modest dynamic range bilaterally. Therefore, no temporary electrode placement was performed. In April 1991 at the age of 20.7 years, she was implanted with a Nucleus multichannel system in the right ear.

The preoperative and postoperative hearing thresholds are shown in Table 2. Even with a powerful hearing aid, no preoperative thresholds could be measured. The implant-aided thresholds after 1 year were slightly better than those of the other patients with Usher's syndrome.

Her results were found to be significantly above chance on the MTS test, at both the syllable and the word recognition level (Fig 1).

Significant scores were obtained on all the pattern recognition tests of the AN battery, except for the Sentence Accent Identification test. No significant trend over testing sessions were seen in these tests. Her scores were significantly above chance on the Vowel Recognition tests, but this was not the case on the Monosyllabic Word Identification and Spondee Identification tests (Fig 1).

A poor CDT performance of about 22 words per minute was found in both the visual-only and auditory-plus-visual conditions as compared to a top score of 60 words per minute

(Fig 2).

Patient D had been using the implant for almost 2 years on a daily basis, for more than 12 hours a day. She could recognize many environmental sounds and had learned to appreciate some kinds of music. She stated that the implant had improved her ability to communicate with other deaf people and with people with normal hearing. She also found that communication in a group of people was improved. She felt more secure and self-confident with the implant and was more optimistic about her future.

Results of Usher's Patients Compared to Other Prelingually Deaf Patients

All the patients compared within this study were profoundly deaf when they entered the selection procedure. The preoperative PTA in decibels HL or the free field average at 0.5, 1 and 2 kHz in decibels HL for all the patients is given in Table 2. The hearing thresholds improved strongly in all the subjects using the implant. No significant difference was found in the implant-aided average thresholds between the group of Usher's patients and the non-Usher's group.

No statistically significant differences were found in the performance of the Usher's group compared to the non-Usher's group on either the syllable recognition level or the word recognition level of the MTS test.

On the tests of the AN test battery, no significant differences were found between the results of the patients with Usher's syndrome and those of the other prelingually deaf on the pattern recognition tests.

Two of the patients with Usher's syndrome and some of the other prelingually deaf patients scored significantly above chance at 1 or more test intervals on the speech recognition tests of the AN battery. Inspection of Fig 1 suggests that patient C, who had a single-channel implant, performed better than the other Usher's patients on the Environmental Sounds Identification test, the Long Vowel Identification test, and on the Monosyllabic Word Identification test; the average score of the other prelingually deaf patients with a single-channel system on the Spondee Identification test was significantly above chance, in contrast to that of the Usher's patients; no significant differences were found between the performance of the two groups of patients on the Short Vowel Identification test.

The postoperative individual CDT scores of the patients with Usher's syndrome are shown in Fig 2. Although the CDT task is not the method of choice for comparing the performances of different patients,[20] the mean scores of the other prelingually deaf patients are also shown in this Figure. Despite their visual handicap, the performance of the patients with Usher's syndrome was not inferior to that of the other patients. In 3 of the 4 Usher's patients and in the other prelingually deaf patients the CDT scores in the auditory-plu-visual and visual-only conditions differed only slightly. No open-set speech recognition in the auditory-only condition was achieved by any of these patients. Similarly, the Usher's patients achieved tracking scores while reading a written text ("top score") that were

comparable to those of the other prelingually deaf patients (means 56 words per minute and 63 words per minute, Student's t=1.17, p>.05).

On the pattern recognition tests of the AN battery, Student-Newman-Keuls grouping showed a significant improvement for the combined group of Usher's and other patients between the average scores at 3 months (52%) and at 12 months (58%) with the 6-month scores in between (54%). The 24-month scores (60%) were not significantly different from the 12-month scores. A trend of improvement was seen on the CDT task in 3 of the patients with Usher's syndrome (Fig 2). However, an analysis of variance on the postoperative MTS scores, speech recognition scores of the AN battery and CDT scores did not show any significant improvement over time.

DISCUSSION

The rehabilitation of the patients with Usher's syndrome included in this study required little extra effort in comparison with that of the other prelingually deaf patients. Adequate illumination and clearly printed testing material were necessary for some of our Usher's patients. They all appeared to be experienced listeners, and the purpose of most of the tests was easily explained to them. They were highly motivated to use oral-aural communication. It should be stressed that all the patients in this study were rehabilitated by a team with extensive experience with teaching prelingually deaf people to optimize the use of their minimal auditory capabilities in oral-aural communication.

Our patients with Usher's syndrome reached significant levels of speech perception as measured with several closed-set tests and their scores did not differ greatly from those of the other prelingually deaf patients. These findings are in agreement with the subjective reports of our patients, who expressed marked improvement in the sound they perceived with a cochlear implant compared to that perceived with a hearing aid. All our patients reported an improvement in communication in everyday life with the implant, as was also described by Eisenberg.[9]

At the pattern recognition level of the AN test battery, a significant improvement was found over time. The 1-year postoperative results were significantly better than the results obtained at the 4-month postoperative testing session. Postoperatively, no significant improvement was found over time at the speech recognition level in the two groups of patients involved in the present study.

No obvious differences were found between the scores from the patients with a single-channel device and the patients with a multichannel device in the Usher's group or in the non-Usher's group. This was underlined by the performance of patient A, who was reimplanted with a multichannel device. On most of the tests she performed as well with the single-channel implant as she did with the multichannel system. Our preliminary findings indicate that prelingually deaf patients might perform equally well with a single-channel system as with a multichannel system. These findings contrast with those in

prelingually deaf children[21] and postlingually deaf adults,[22] who demonstrated superior auditory performance with a multichannel implant. Single-channel implants using analogue coding strategies can transmit the temporal aspects of sound very effectively; obviously this allows for the transmission of environmental sounds and many prosodic aspects of speech.[23] The good signal function of single-channel implants is of great importance to the prelingually deaf. Furthermore, single-channel implants could be less costly than multichannel implants and the fitting of analogue systems is much simpler in general.

The majority of patients with Usher's syndrome suffer from progressive visual impairment, which leads to "tunnel vision" and night blindness. If the level of sound and speech pattern recognition achieved in the present patients can be maintained even after they become blind, a cochlear implant will help them to feel safer, more independent and less isolated.

CONCLUSIONS

Rehabilitation of implanted patients with Usher's syndrome using oral-aural communication required very little extra effort compared to the rehabilitation of other prelingually deaf patients.

After implantation, adult patients with Usher's syndrome obtained significant scores on closed-set speech recognition tests, which were comparable to the results of other prelingually deaf adults. No open-set speech recognition was possible in any of our patients presented here. Results of the Usher's patients with a Nucleus multichannel implant were not significantly better than those of the Usher's and other prelingually deaf patients with a single-channel system.

The positive subjective experience and the significantly improved testscores justify the continuation of the implantation of carefully selected prelingually deaf candidates. However, it remains important to adjust expectations to a realistic level.

REFERENCES

1. Gantz BJ, Tye Murray N, Tyler RS. Word recognition performance with single-channel and multichannel cochlear implants. Am J Otol 1989;10:91-4.
2. Dowell RC, Mecklenburg DJ, Clark GM. Speech recognition for 40 patients receiving multichannel cochlear implants. Arch Otolaryngol Head Neck Surg 1986;112:1054-59.
3. Berliner KI, Luxford WM, House WF. Cochlear Implants: 1981-1985. Am J Otol 1985;2:173-86.
4. Clark GM, Busby PA, Roberts SA Dowell RC, Blamey PJ, Mecklenburg DJ, Webb RL, Pyman BC, Burkhard KF. Preliminary results for the cochlear corporation multielectrode intracochlear implant in six prelingually deaf patients. Am J Otol 1987;8:234-9.
5. Tong YC, Busby PA, Clark GM. Perceptual studies on cochlear implant patients with early onset of profound hearing impairment prior to normal development of auditory, speech and language skills. J Acoust Soc Am 1988;84:951-62.
6. Dawson PW, Blamey PJ, Rowland LC, Dettman SJ, Clark GM, Busby PA, Brown AM, Dowell RC, Rickards FW. Cochlear implants in children, adolescents, and prelinguistically deafened adults: speech perception. J Speech Hearing Res 1992;35:401-417.
7. Waltzman SB, Cohen NL, Shapiro WH. Use of a multichannel cochlear implant in the congenitally and prelingually deaf population. Laryngoscope 1992;102:395-399.
8. Vernon M. Usher's syndrome: deafness and progressive blindness. Clinical cases, prevention, theory and literature survey. J Chron Dis 1969;22:133-151.
9. Eisenberg LS. Use of the cochlear implant by the prelingually deaf. Ann Otol Rhinol Laryngol 1982;91(suppl 91):62-66.
10. Martin EL. Speech recognition by a deaf-blind multichannel cochlear implant patient. Ear Hear 1988;9:70-74.
11. van den Broek P, Admiraal R. Selection of patients for a cochlear implant. Acta Otorhinolaryngol Belg 1991;45:243-246.
12. van den Broek P, Coninx F, Mens L, Admiraal R, Brokx J. Temporary placement of electrodes as part of a selection procedure for cochlear implants. In: Charachon R, Garcia Ibanez E, eds. Long term results and indications in otology and otoneurosurgery. POL 051-3. Proceedings of the Politzer Conference in Ibiza, 1989 and Courchevel, 1990. Amsterdam, the Netherlands: Kugler Publications, 1990:1-3.
13. Erber HP, Alencewicz CM. Audiologic evaluation of deaf children. J Speech Hear Dis 1976;41:256-267.
14. Brokx JPL, Beynon AJ, van Durme M, Timmermans B. Ontwikkeling van Nederlandstalige auditieve tests. Logopedie en Foniatrie 1987;6:186-190.
15. Beynon AJ, Brokx JPL, Mens LHM, van den Broek P. De eerste resultaten met een elektrische binnenoorprothese. Logopedie en Foniatrie 1990;62:113-116.
16. Owens E, Kessler D, Raggio M. Analysis and revision of the Minimal Auditory Capabilities (MAC) battery. Ear Hear 1985;6:80-290.
17. Tyler RS, Preece JP, Lowder MW. The Iowa cochlear implant tests; university school of medicine, Iowa city, Iowa: Department of Otolaryngology-Head and Neck Surgery, University of Iowa, 1983.
18. De Filippo CL, Scott BL. A method for training and evaluating the reception of ongoing speech. J Acoust Soc Am 1987;63:1186-92.
19. Morgan DE, Dirks DD, Bower DR. Suggested threshold sound pressure levels for frequency modulated (warble) tones in the sound field. J Speech Hear Dis 1972;44:37-54.
20. Tye-Murray N, Tyler RS. A critique of continuous discourse tracking as a test procedure. J Speech Hear Dis 1988;53:226-231.

21. Miyamoto RT, Osberger MJ, Robbins AM, Myres WA, Kessler K, Pope MC. Longitudinal evaluation of communication skills of children with single- or multichannel cochlear implants. Am J Otol 1992;13:215-222.
22. Doyle PJ, Pijl S, Noel FJ. The cochlear implant: a comparison of single-channel and multichannel results. J Otolaryngol 1991;20:204-208.
23. Tyler RS, Moore BC, Kuk FK. Performance of some of the better cochlear implant patients. J Speech Hear Res 1989;32:887-911.

CHAPTER FIVE

PERFORMANCE OF PRELINGUALLY VERSUS POSTLINGUALLY DEAFENED PATIENTS USING SINGLE OR MULTICHANNEL COCHLEAR IMPLANTS

Johannes B. Hinderink, MD, Lucas H.M. Mens PhD,
Jan P.L. Brokx, PhD, Paul van den Broek, MD, PhD

Published in slightly abbreviated form in *Laryngoscope* 105:1995 pp. 618-622

PERFORMANCE OF PRELINGUALLY VERSUS POSTLINGUALLY DEAFENED PATIENTS USING SINGLE OR MULTICHANNEL COCHLEAR IMPLANTS

Johannes B. Hinderink, MD, Lucas H.M. Mens PhD,
Jan P.L. Brokx, PhD, Paul van den Broek, MD, PhD

ABSTRACT

The auditory and aided lip-reading performance of 8 prelingually and 11 postlingually deaf patients who had received a single or multichannel cochlear implant was evaluated during 2 years follow-up. The auditory performance was investigated using closed-set pattern and speech recognition tests and a Continuous Discourse Tracking (CDT) task. Although all the patients improved on the pattern recognition level, the most significant improvement was observed in the group of postlinguals who were using a multichannel implant. Only small differences were found between the prelinguals who were using a single or multichannel system and the postlinguals who were using a single-channel system. Comparable results were found on the speech recognition level but the outstanding performance of the postlinguals who were using a multichannel system was even more pronounced. The results on a Continuous Discourse Tracking task were similar; furthermore, speech recognition in the auditory-only condition was only achieved by the postlinguals who were using a multichannel system. On average, the users' evaluations obtained by means of a questionnaire were positive in all the different user groups. It was concluded that it is feasible to implant highly motivated patients who became deaf prelingually and have learnt to use oral-aural communication. Single-channel systems may be as effective as multichannel systems in this group.

INTRODUCTION

The efficacy and safety of cochlear implantation in postlingually deaf adults has been well-established by numerous studies,[1 2 3] including a large prospective randomized trial [4] In general, the performance with multichannel devices is superior to that with single-channel devices in this group of patients, especially for understanding speech

Similar results have also been found in post and prelingually deaf children [5 6] Multichannel users appeared to have higher performance levels and faster rates of learning than single-channel users Whereas the performance of the single-channel users reached a plateau after 1 5 years, the multichannel users continued to show improvement after 2 or more years and a considerable number of them could understand speech in open-set tests

In contrast, only a few studies have specifically focused on the results of implantation in prelingually deaf adults No direct comparisons have been made between the results of different cochlear implant systems in this group of adults

Eisenberg[7] reported on the subjective benefits with a single-channel system in 12 adults with various communication backgrounds (manual, signing or both) who had become deaf prelingually Eight of them were considered to be successful users, who were able to respond to environmental sounds and reported feeling more independent, more comfortable socially and less lonely

Tong, Busby and co-workers[8 11] compared the psychophysical and speech perception performance in a small number (n#4) of prelingually deaf adults and one adolescent to the performance of a group of postlingually deaf adults who were implanted with the Nucleus multichannel device Although there was considerable variation in performance among the prelingually deaf patients, their performance on speech perception tests was inferior to that of the postlinguals The performance of the prelinguals on several psychophysical tests was similar to that of the postlinguals, but their performance on electrode confusion tests, which reflect the processing of tonotopic information, was inferior

Burian[12] reported on 6 prelingually deaf adults who had been implanted with the single-channel Vienna system Half of them showed some improvement in the lip-reading-plus-implant condition, but the other 3 only benefited with regard to the perception of environmental sounds

A limited number of studies, each involving only a small number (n#3) of prelingually deaf adults who were implanted with the Nucleus multichannel system,[13 14 15] reported significant improvements on speech perception tests, with or without lip-reading and also mentioned subjective benefit

In the present study, the speech recognition performance of a group of

Patient No.*	Sex	Etiology	Age at onset of deafness (yrs)	Duration of deafness (yrs)	Age at implantation (yrs)	Implant system†	Ages at which hearing aids were used (yrs)
ES4/ EM221‡	F	Usher	0	20.1/23.2	20.1/23.2	S/M	20
ES5	M	Meningitis	0.3	20.5	20.8	S	
ES7	M	Meningitis	1.6	22.3	23.8	S	
ES10	M	Meningitis	2.3	17.3	19.5	S	
ES12	M	Usher	0	28.9	28.9	S	
ES29	F	Mondini	0	24.3	24.3	S	
EM23	F	Usher	0	21.0	21.0	M	20
EM30	M	Hereditary	0	33.5	33.5	M	20
OS1	M	Trauma	8.3	20.5	28.8	S	
OS2	M	Progressive	31.6	5.0	36.6	S	
OS3	M	Meningitis	10.8	47.8	58.6	S	
OS8	F	Meningitis	10.7	45.5	56.2	S	
OM6	F	Meningitis	10.8	43.7	54.4	M	15
OM13	M	Meningitis	6.2	20.2	26.3	M	20
OM14	M	Meningitis	37.3	5.9	43.2	M	20
OM15	F	Unknown	26.3	36.0	62.3	M	20
OM16	F	Meningitis	14.3	23.1	37.3	M	18
OM19	F	Otosclerosis	44	16.3	60.3	M	16
OM21	M	Meningitis	7.4	43.9	51.3	M	18

*The letters in the patient identification code bear the following meanings: O = postlingually deaf; E = prelingually deaf; S = single-channel cochlear implant system; M = multi-channel cochlear implant system. The number is the implant program patient identification number.
†Single-channel (S) cochlear implant or multichannel (FM) cochlear implant.
‡This prelingually deaf patient used a single-channel system for two years and then was reimplanted with a multichannel system.

Table 1. Patient data for 19 deaf adults who received cochlear implants

postlingually deaf adults who were using either a single-channel or a multichannel cochlear implant was compared to that of a group of prelingually deaf adults who were using the same types of implants. Speech recognition tests were performed at various intervals during a follow-up period of 2 years to see whether the prelinguals benefitted significantly from the implantation, and to see to what extent this benefit differed for the users of a single-channel versus a multichannel system. Additionally, the subjective experience of all the patients was evaluated by means of a questionnaire.

PATIENTS AND METHODS

Patients

Some characteristics of the 19 adults who were implanted in the Nijmegen/St

Michielsgestel implant programme between 1988 and 1991 are given in Table 1. The subjects were assigned an identification code which began with O or E (O for the postlinguals, E for the prelinguals), followed by S or M (S for a single channel, M for a multichannel cochlear implant) and the implant programme identification number. One of the prelinguals (ES4) was reimplanted with a Nucleus multichannel system (EM22) after two years of using a single-channel implant and was monitored for the subsequent two years. The mean age of the prelingually deaf patients (n=8) and the postlingually deaf patients (n=11) at the time of implantation was 24 and 47 years, respectively. The mean duration of deafness was 24 years for the prelinguals and 28 years for the postlinguals. All the prelinguals were (former) pupils at the Institute for the Deaf in St. Michielsgestel and had completed oral-aural communication programmes. All the patients came from middle-class backgrounds and had normal intelligence. They all had profound bilateral deafness and had experienced no benefit from a hearing aid. A full description of the selection procedure has been given elsewhere.[16,17] Post-operative tests were performed at 3, 6, 12 and 24 months follow-up on all of the patients, except for patient ES7, who stopped using his implant during the first year. For the presentation of the results, four subgroups of patients were distinguished, namely the prelingually deaf with a single or a multichannel system and the postlingually deaf with a single or a multichannel system.

Implantation

The single-channel systems (Med-El\E1 or 3M/Vienna) were implanted with the active electrode placed extracochlearly in the round window niche and the reference electrode inserted into the bone superior to the lateral semicircular canal. In all but 4 of the users of the Nucleus Mini System 22 (listed in Table I), all the electrodes could be placed succesfully in the scala tympani.

Rehabilitation

The processors were fitted after a post-operative recovery period of at least four weeks. An MSP speech processor with the MPEAK coding scheme was used for the Nucleus system. The patients and their rehabilitation partners were then admitted to the Institute for the Deaf for a period of two weeks to take part in the initial phase of the rehabilitation programme. Every two months during the first year of follow-up, the patients participated in a one day training and testing session at the Institute for the Deaf. Occasional processor readjustments were performed at the University Hospital Nijmegen. Processor adjustment tended to be prolonged beyond the first rehabilitation year for some of the prelingually deaf users.

The evaluative tests were administered pre-operatively and 3, 6, 12 and 24 months after the processor had been fitted. The pre-operative test results in the auditory-only modality were not significantly above chance for any of the patients, therefore only the

post-operative results were analysed.

Test materials

The auditory perception of segmental and suprasegmental aspects of speech was assessed by means of a Dutch version of the Monosyllable-Trochee-Spondee (MTS) test[18] and by means of the Antwerpen-Nijmegen (AN) test battery.[19] The AN test battery has a similar set up to the MAC test battery[20] and it comprises tests for speech recognition (i.a. Short Vowel Identification, Long Vowel Identification, Monosyllabic Word Identification and Spondee Identification), pattern recognition (i.a. Number of Syllables, Sentence Accent Identification, Male/Female/Child Voice Discrimination) and an Environmental Sounds Identification test. All the AN tests are in a closed-set format.

Lip-reading skills were tested using Continuous Discourse Tracking (CDT)[21] in the visual-only condition (V) and in the auditory-plus-visual condition (AV). If possible, the CDT task was also performed in the auditory-only condition (A).

Additionally, the users' evaluations were recorded by means of the "Gestel-Nijmegen implant questionnaire" at one year follow-up. The questions concerned 4 categories, namely (1) implant use, (2) communication, (3) feeling of safety, and (4) satisfaction with the implant. The most important questions are given in the appendix.

The AN tests were administered in a double-walled sound booth. The cassette recordings of the AN tests were presented by means of a tape recorder coupled to the standard audiometer (Interacoustics AC5) with a free-field amplifier. The tests were presented at a comfortable volume level which was determined at the beginning of each session using extra examples of the test items. The loudspeaker was placed one meter in front of the patient.

The MTS and the CDT tests were administered in a quiet room and were spoken live by one of the rehabilitation therapists who was sitting approximately 1 m away from the patient.

During all the tests, the sensitivity of the speech processor was set at the same level as in normal daily use.

Data analysis

Analysis of variance (General Linear Models procedure, SAS Institute) was used to evaluate the different test scores on the closed-set tests. Independent factors were: Age at Onset (prelingually deaf or postlingually deaf), Implant System (single channel or multichannel), Test Type (the MTS and individual AN tests) and Follow-up Interval (3, 6, 12 and 24 months after the processor had been fitted). The patients formed random variables, nested in Age at Onset and Implant System. The ANOVA was limited to the main effects and the first-order interactions due to the limited number of observations. A separate analysis of variance was performed on the scores on the CDT task. Post-hoc

Student Newman Keuls' multiple range tests (alpha=5%, GLM) were performed on the adjusted scores for each follow-up interval, averaged across all the patients.

For analysis, the scores on the closed-set pattern and speech recognition tests were adjusted for the number of test items and the number of alternatives, using the following formula:

$$S_{ad} = \{(C-I/(N-1))/n\} \times 100$$

S_{ad} =adjusted score, C=number of correct items, I=number of incorrect items, N=number of alternatives, n=total number of items.

To analyse the performance on the pattern recognition level, the adjusted scores on the pattern recognition tests of the AN test battery (Number of Syllables Identification, Sentence Accent Identification, Male/Female/Child Voice Discrimination and Environmental Sounds Identification test) and on the MTS pattern recognition test were pooled. The same was done for the speech recognition level, using the speech recognition tests of the AN battery (Short Vowel Identification, Long Vowel Identification, Monosyllabic Word Identification and Spondee Identification test), and the MTS speech recognition test. The pooled adjusted scores are referred to as "composite scores."

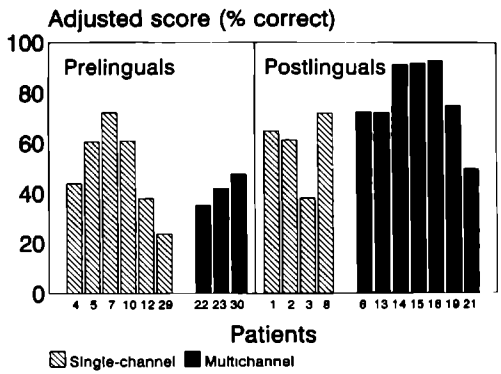

Figure 1. Composites of chance adjusted scores on the five pattern recognition tests at 12 months follow-up for the individual patients. Patients are grouped according to age at onset of deafness and implant system.

RESULTS

Pattern recognition

The average pattern recognition composite scores at 1 year follow-up are shown in Figure 1 for the individual patients, grouped according to the four subgroups. Inspection of this figure reveals that there were no great differences on the pattern recognition level between the various groups of patients.

Significant differences were found between the adjusted scores on the various tests [$F(1,16) = 6.63$, $p = 0.02$] which indicates that some of the tests were more difficult than

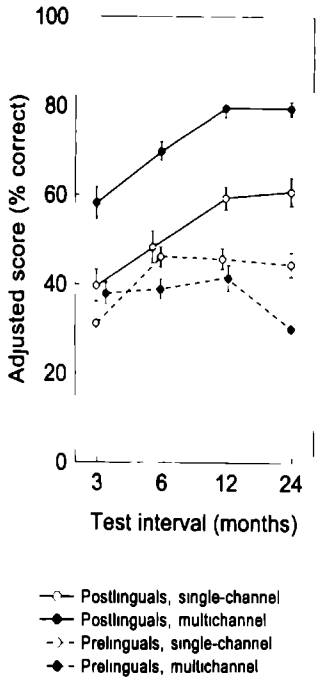

Figure 2. Composite scores on the pattern recognition tests for the four groups of patients at 3, 6, 12, and 24 months after cochlear implantation. Vertical bars represent 1 standard error.

others but Test Type did not interact with any of the other variables.

Figure 2 shows the composite scores averaged over Test Type for the four groups of patients at the various follow-up intervals. The average performance improved significantly over time [$F(3,45) = 6.93$, $p = 0.0006$]. Post-hoc SNK grouping revealed that the average performance improved significantly between 3 and 6 months, but not after the 6 months follow-up. The postlingually deaf users performed better than the prelingually deaf users [$F(1,16) = 19.49$, $p = 0.0004$]. The improvement over time differed between the pre and postlingually deaf [$F(3,45) = 3.16$, $p = 0.03$]: inspection of Figure 2 shows greater and more prolonged improvement over time for the postlinguals. No other significant effects were found.

Speech recognition

Figure 3 shows the composite scores for each patient on the speech recognition level at 1 year follow-up. In contrast to the pattern recognition level, a substantial difference can be seen between the performance of the postlingually deaf who were using the multichannel system and that of the rest of other patients.

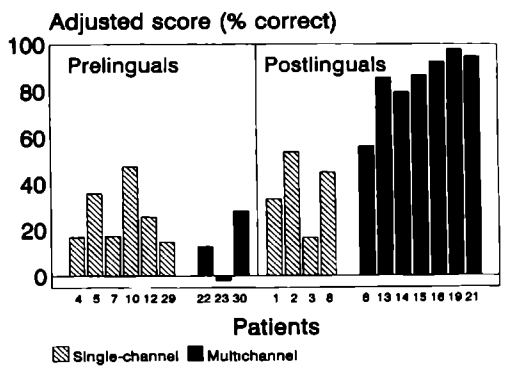

Figure 3. Composite scores on the 5 speech perception tests at 12 months follow-up for the individual patients. The patients are grouped according to age at onset of deafness and implant system.

As Test Type interacted significantly with both Age at Onset [$F(4,64) = 4.21$, $p = 0.004$], and Implant System [$F(4,64) = 5.29$, $p = 0.001$], the test scores for each of the four groups of patients are shown in Figure 4 for the individual speech recognition tests. It should be noted that the raw non-adjusted scores are presented in this Figure.

From Figure 4 it is evident that these interactions can be ascribed to the performance of the postlinguals with a multichannel

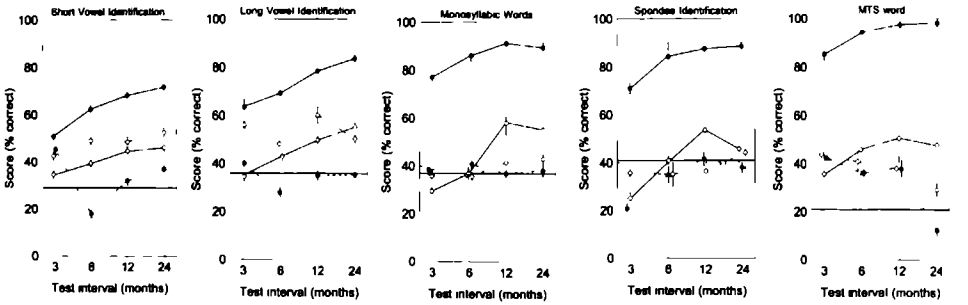

Figure 4. Test scores on the individual speech perception tests for the four groups of patients. The full lines indicate the upper limit of the 95% confidence interval. Vertical bars represent 1 standard error. Note that the raw unadjusted scores are presented in this figure (MTS = Monosyllable-Trochee-Spondee test).

implant.

Figure 5 shows the speech recognition composite scores for the four groups of patients during follow-up. The mean performance improved significantly over time [$F(3,45) = 7.20$, $p = 0.0005$]. SNK grouping of the means of all patients showed that the performance improved significantly between 3 and 6 months follow-up and between 6 and 12 months follow-up. The improvement between 12 and 24 months follow-up was not significant. As can be seen in Figure 5, the improvement over time was observed mainly in the group of postlingually deaf patients, which was substantiated by the finding of an interaction between Age at Onset and Follow-up Interval [$F(3,45) = 3.84$, $p = 0.02$].

A multichannel implant was of significantly greater benefit than a single-channel implant for speech recognition only in the group of postlingually deaf [$F(1,16) = 15.00$, $p = 0.001$].

Continuous Discourse Tracking

The auditory-plus-visual (AV) enhancement over the visual-only performance in the CDT task was determined as follows: ((AV-V)/V)x100. The postlingually deaf users showed greater AV enhancement than the prelingually deaf users [$F(1,16) = 5.63$, $p = 0.03$]. Figure 6 shows that the enhancement in the postlinguals mainly occured in the group fitted with a multichannel implant [$F(1,16) = 4.89$, $p = 0.04$]. Post-

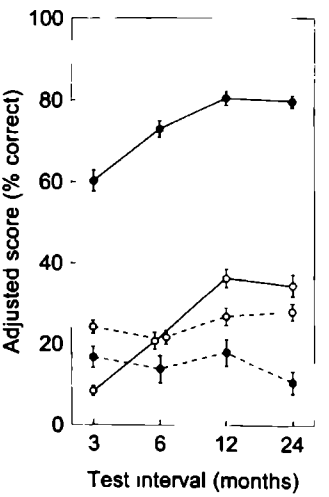

Figure 5. Composite scores on the speech recognition level for the four groups of patients at 3, 6, 12 and 24 months follow-up. Bars represent 1 standard error.

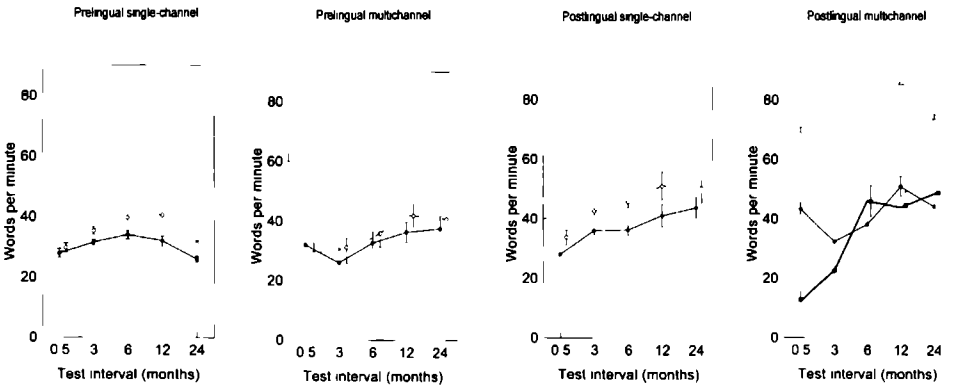

Figure 6. Continuous Discourse Tracking scores for the different groups of patients. The dotted lines represent the auditory-plus-visual scores, the solid lines the visual-only scores and the thick line the auditory-only scores. Bars represent 1 standard error

hoc analysis of variance on the AV and V scores of the post and prelingually deaf revealed that the two groups of patients with a single-channel implant experienced significant benefit in the AV condition over the V condition: [$F(1,3) = 11.89$, $p = 0.04$] and [$F(1,5) = 6.97$, $p = 0.05$] respectively. No significant enhancement was found in the group of prelinguals with the Nucleus system. There was no significant increase of the enhancement over time in any of the groups.

Speech recognition in the auditory-only condition (A) was only achieved by the postlingually deaf with a multichannel system as is shown in Figure 6 (thick grey line). This performance improved significantly over time [$F(4,33) = 7.84$, $p = 0.0004$] and SNK grouping showed significant improvement between the 3 and 6 months follow-up, but the performance did not improve significantly at the 5% level.

Subjective benefits: the GN implant questionnaire at 1 year follow-up

Each category is discussed individually.

(1) Implant use: 17 out of the 19 patients in this study were using their implant in all situations. One patient only used the implant in some situations (OS1) and 1 patient (ES7) stopped using the implant one year after implantation, because of unsatisfactory sound detection and subjective annoying and sometimes painful sounds.

(2) Communication: 17 out of the 19 patients reported that communicating with one other person had become easier by using the implant. Two patients, ES5 and ES7, reported no changes. Ten out of the 19 patients felt more at ease when communicating with other deaf people, while 16 out of the 19 patients reported feeling more relaxed when communicating with people with normal hearing and 2 patients (ES5, ES7) felt more tense while using their implant.

(3) Feeling of safety: 9 out of the 11 postlinguals and 7 out of the 8 prelinguals, including all the patients with the Usher syndrome, reported feeling more safer in traffic while using the implant. Only one patient (ES29) felt less secure because of annoying traffic sounds.

(4) Satisfaction: 16 out of the 19 patients were content with the implant, 2 (OS3 and ES12) were somewhat disappointed and only one patient (ES7) was greatly disappointed with the result and had stopped using his implant.

DISCUSSION

Despite the fact that this study involved a limited number of patients, some preliminary conclusions can be drawn from the results.

The performance of the postlingually deaf on the pattern recognition level was found to be superior to the performance of the prelingually deaf, which is in accordance with previous studies.[8 9 12] Nevertheless, the average performance of the prelinguals on the pattern recognition level was significantly above chance. The type of implant used had no significant effect on the results of the pattern recognition tests.

On the closed-set speech recognition tests, the performance of the postlinguals with a multichannel system was clearly superior to that of the postlinguals with a single-channel system and to that of the two groups of prelinguals. No significant difference in performance was found between the postlinguals with a single-channel system and the prelinguals with either system. In the prelinguals, no significant differences were found with regard to the type of implant used. Again, however, the average performance of the prelinguals was found to be significantly above chance.

Although comparison of the CDT scores from the different individuals has limited value because of the semi-standardized presentation of the test materials and possible differences between the presentation of the test,[22] the AV enhancement was found to be largest in the postlinguals with a multichannel implant, which is in line with the results on the pre-recorded closed-set speech recognition tests. Furthermore, the postlinguals with a multichannel implant were the only patients who achieved open-set speech recognition in the auditory-only condition. However, a small but statistically significant benefit was found in the auditory-plus-visual condition compared to the visual-only condition in the post and prelinguals with a single-channel system, but not in the present group of prelinguals with a multichannel system. It should be noted that the latter group was too small to draw definitive conclusions.

The benefit experienced by the postlinguals with the multichannel system became especially apparant during the speech recognition tests, such as the Monosyllabic Word and Spondee Identification tests, the MTS speech recognition test and the CDT task. Their better performance could be attributed to the pre-existing language skills. Their

superior performance was much less evident on the Short and Long Vowel Identification tests. This is in line with the findings on the pattern recognition level described above.

Despite the only moderate results of the prelingually deaf, almost all of them reported that they were satisfied with their implant and they claimed that it had added quality to their life. Only one of the prelinguals stopped using the implant because of persistent annoying and sometimes painful sounds, despite several readjustments. It is interesting to note that this was the only prelingual in our study who found a deaf partner after he had been implantated. He subsequently started to lean more and more towards the deaf community, whereas the other prelinguals in our study were highly motivated to continue using oral-aural communication and were becoming increasingly involved in the "hearing" world.

Several studies have shown that postlingually deaf adults benefited more from a multichannel implant than from a single-channel implant, although the patients who performed best in each group achieved a comparable level of (open-set) speech recognition.[25] In the present study, we did not encounter any patients with such an outstanding performance.

Contrary to previous investigations, this study compared different types of implant in a group of prelinguals. Some of these studies have shown that prelinguals do benefit from implantation with either a multichannel or a single-channel implant. This is in agreement with the present findings on the closed-set auditory tests and even on the more or less open-set CDT task. Our findings suggest that prelingually deaf adults can achieve similar performance using either an extra-cochlear single or an intracochlear multichannel implant. This conclusion was confirmed by the performance of patient ES4/EM22 who was reimplanted with a Nucleus multichannel system after failure of her single-channel system.

It is important to note that the benefit of implantation should not be expressed only by enhanced performance on speech recognition tests. Personal factors, such as ease of communication with other people, stress reduction when participating in traffic, or the psychological benefit of actually being able to hear sounds, are difficult to assess objectively by means of tests. The results of the questionnaire in this study indicate that these factors play an important role in the amount of benefit provided by the implant.

It is open to discussion whether the present results can justify the large-scale implantation of prelingually deaf adults. In this study all exept for one of the prelinguals showed a persistent high motivation to learn to function in the "hearing world" by means of oral-aural communication. The present results indicate that it is feasible to implant highly motivated, oral-aural oriented patients who became deaf prelingually, but it is doubtful whether any additional benefit can be gained from implanting highly sophisticated expensive multichannel systems in this group of patients.

REFERENCES

1. Gantz, B.J., Tye Murray, N. and Tyler, R.S.: Word recognition performance with single-channel and multichannel cochlear implants. Am J Otol 1989;10:91-94.
2. Dowell, R.C., Mecklenburg, D.J. and Clark, G.M.: Speech recognition for 40 patients receiving multichannel cochlear implants. Arch Otolaryngol Head Neck Surg 1986;112:1054-1059.
3. Berliner, K.I., Luxford W.M. and House, W.F.: Cochlear Implants: 1981-1985. Am J Otol 1985;2:173-186.
4. Cohen, N.L., Waltzman, S.B., Fischer, S.G., et al.: A prospective, randomized study of cochlear implants. New Eng J Med 1993;328:233-237.
5. Staller, S.J., Dowell, R.C., Beiter, A.L., et al.: Perceptual abilities of children with the Nucleus 22-channel cochlear implant. Ear Hear 1991;12Supp:34-47.
6. Miyamoto, R.T., Osberger, M.J., Robbins, A.M., et al.: Longitudinal evaluation of communication skills of children with single- or multichannel cochlear implants. Am J Otol 1992;13:215-222.
7. Eisenberg, L.S.: Use of the cochlear implant by the prelingually deaf. Ann Otol Rhinol Laryngol 1982;91:62-66.
8. Tong, Y.C., Busby, P.A. and Clark, G.M.: Perceptual studies on cochlear implant patients with early onset of profound hearing impairment prior to normal development of auditory, speech and language skills. J Acoust Soc Am 1988;84:951-62.
9. Busby, P.A., Roberts, S.A., Tong, Y.C., et al.: Results of speech perception and speech production training for three prelingually deaf patients using a multiple-electrode cochlear implant. British J Audiol 1991;25:291-302.
10. Busby, P.A., Tong, Y.C. and Clark, G.M.: Psychophysical studies using a multiple-electrode cochlear implant in patients who were deafened early in life. Audiology 1992;31:95-111.
11. Busby, P.A., Tong, Y.C. and Clark, G.M.: Electrode position, repetition rate, and speech perception by early- and late-deafened cochlear implant patients. J. Acoust. Soc. Am. 1993;93:1058-1067.
12. Burian, K.: Derzeitiger stand der elektrostimulation der hornervenfasern mit cochlearimplantaten. Wiener Medizinischen Wochenschrift 1992;11/12:264-273.
13. Dawson, P.W., Blamey, P.J., Rowland, L.C., et al.: Cochlear implants in children, adolescents, and prelinguistically deafened adults: speech perception. J Speech Hearing Res 1992; 35:401-417
14. Waltzman, S.B., Cohen, N.L. and Shapiro, W.H.: Use of a multichannel cochlear implant in the congenitally and prelingually deaf population. Laryngoscope 1992;102:395-399.
15. Skinner, M.W., Binzer, S.M., Fears, B.T., et al.: Study of the performance of four prelinguistically deaf patients with a multi-electrode, intracochlear implant. Laryngoscope 1992;102:797-806.
16. Broek, P. van den, Coninx, F., Mens, L., et al.: Temporary placement of electrodes as part of a selection procedure for cochlear implants. In: Charachon R. and Garcia Ibanez E. eds. Long term results and indications in otology and otoneurosurgery. POL 051-3. Proceedings of the Politzer Society Conference in Ibiza, 1989 Courchevel, 1990. Amsterdam, the Netherlands: Kugler publications 1990;1-3.
17. Broek, P. van den and Admiraal, R.: Selection of patients for a cochlear implant. Acta Oto-rhino-laryngol Belg 1991;45:243-246.
18. Erber, H.P. and Alencewicz, C.M.: Audiologic evaluation of deaf children. J Speech Hear Dis 1976;41:256-267.

19 Brokx, J PL , Beynon, A J , Durme, M van, et al Ontwikkeling van Nederlandstalige auditieve tests Logopedie en Foniatrie 1987,59 186-190

20 Owens, E , Kessler D and Raggio, M Analysis and revision of the Minimal Auditory Capabilities (MAC) battery Ear Hear 1985,6 280-290

21 De Filippo, C L and Scott, B L A method for training and evaluating the reception of ongoing speech J Acoust Soc Am 1987,63 1186-1192

22 Tye-Murray, N and Tyler, R S A critique of continuous discourse tracking as a test procedure J Speech Hear Dis 1988,53 226-231

23 Tyler, R S , Moore, B C and Kuk, F K Performance of some of the better cochlear implant patients J Speech Hear Res 1989,32 887-911

APPENDIX GN IMPLANT QUESTIONNAIRE

Question 4

I wear my cochlear implant

1 in all situations
2 in a few situations
3 never

Question 5

I use my speech processor

1 > 12 hours/day
2 3-12 hours/day
3 < 3 hours/day

Question 10

Owing to the cochlear implant, I have

1 fewer problems with communication
2 more problems with communication
3 not experienced any change

Question 11 (you may fill in more than one answer)

Owing to the cochlear implant, I have

1 more contact with people with normal hearing
2 more contact with the deaf and hearing impaired
3 less contact with people with normal hearing
4 less contact with the deaf and hearing impaired
5 not experienced any change

Question 14

Owing to the cochlear implant, I have

1 more frequent contact with the deaf and hearing impaired
2 less frequent contact with the deaf and hearing impaired
3 the same amount of contact as before the implant

Question 15

Owing to the cochlear implant, I have

1 more frequent contact with people with normal hearing
2 less frequent contact with people with normal hearing
3 the same amount of contact as before the implant

Question 19

Using my cochlear implant to communicate with others who are deaf or hearing impaired, I feel

1 more relaxed than I did before the implant
2 less relaxed than I did before the implant
3 the same as before the implant

Question 20

Using my cochlear implant to communicate with others who have normal hearing, I feel

1 less relaxed than I did before the implant
2 more relaxed than I did before the implant
3 the same as before the implant

Question 26 (you may fill in more than one answer)

While communicating with people with normal hearing, the cochlear implant has meant that

1 there is less need to use written communication
2 there is less need to lip-read
3 lip-reading is easier
4 there is less need for sign language
5 nothing has changed

Question 46

Using my cochlear implant out of doors, I feel

1 less safe than I did without the implant
2 safer than I did without the implant
3 the same as without the implant

Question 49

My speech while using the cochlear implant has

1 become easier
2 become more difficult
3 not changed

Question 50

Using the cochlear implant, others have found that my speech is

1 easier to understand
2 more difficult to understand
3 the same as it was before the implant

Question 53

Having a conversation with 1 other person while using the cochlear implant is

1 usually more difficult than before the implant
2 usually easier than before the implant
3 generally unchanged

Question 69

My results achieved with the cochlear implant are

1 very satisfactory
2 fairly satisfactory
3 unsatisfactory
4 very unsatisfactory

Question 49

Are you disappointed with the results you have achieved with the cochlear implant?

1 yes
2 a little
3 no

Question 74

The cochlear implant has meant that my life is

1 much more pleasant
2 a little more pleasant
3 a lot less pleasant
4 slightly less pleasant

Question 75

Has the cochlear implant led to improvements in your way of life?

1 to some extent
2 no
3 yes

CHAPTER SIX

DEVELOPMENT AND APPLICATION OF A HEALTH-RELATED QUALITY OF LIFE INSTRUMENT FOR ADULTS WITH COCHLEAR IMPLANTS: THE NIJMEGEN COCHLEAR IMPLANT QUESTIONNAIRE

Johannes B. Hinderink, MD; Paul F.M. Krabbe, PhD;
Paul van den Broek, MD, PhD

Otolaryngology Head and Neck Surgery 123: 2000 pp. 756-765

DEVELOPMENT AND APPLICATION OF A HEALTH-RELATED QUALITY OF LIFE INSTRUMENT FOR ADULTS WITH COCHLEAR IMPLANTS: THE NIJMEGEN COCHLEAR IMPLANT QUESTIONNAIRE

Johannes B. Hinderink, MD; Paul F.M. Krabbe, PhD;
Paul van den Broek, MD, PhD

ABSTRACT

Objective To develop a quantifiable, self-assessment health-related quality of life (HRQoL) instrument for use in cochlear implant (CI) users

Design Three principal domains were distinguished physical, psychological and social Forty-five postlingually deaf adult multichannel cochlear implant users and forty-six deaf candidates on the waiting list for a CI (control group) participated in the study

Results Retrospective scores for the CI group corresponded very well with the scores for the control group Current QoL scores were substantially higher for all six subdomains Internal consistency and test-retest reliability coefficients proved to be satisfactory while the ability to detect clinical changes with the NCIQ proved to be good

Conclusions The psychometric characteristics of the NCIQ proved to be reliable, probably valid and sensitive to clinical changes The data obtained with the NCIQ reflected that the instrument was able to detect that a CI had significant effects on several HRQoL aspects, including the social and psychological domains

INTRODUCTION

Cochlear implantation (CI) is a well-accepted and routinely used treatment for profound deafness.[1] Objective speech perception results have improved over the past decades by using advanced technology in this field.[2] Today most patients achieve a certain degree of open-set speech understanding with or without lip-reading. Apart from the improvement in hearing, important improvements may also be expected on other aspects.

Clinicians and policy makers have recognized that changes in a patient's quality of life (or health status) are the primary outcome of medical interventions. In the case of CI this means that the treatment not only affects hearing and speech production, but it also has an impact on self-esteem, daily activities and social functioning. Health outcomes on the subjective level of the patient can be measured in terms of "health-related quality of life" (HRQoL).

In a review of the literature we found that studies investigating the impact of CI have less scope than studies reporting specific benefits measured by sound and speech perception tests. Most of the studies that focused on CI-HRQoL were generally based on the use of open-ended questionnaires or on interviews with patients.[3-5] Although these types of study may yield valuable information, the manner of data collection prevents us from making any systematical comparisons between groups of patients or evaluating follow-up outcomes. A few authors made use of closed-set questionnaires that produced quantifiable scores. The most elaborate of these studies was conducted by Maillet et al.[6] who used three questionnaires to measure changes in the quality of life of CI users: the Patient Quality of Life Form, the Index Relative Questionnaire Form to evaluate the opinions of the patient's relative (e.g. partner, parent) and the Performance Inventory for Profound Hearing Loss Answer Form. The latter can also be used for hearing impaired people. Although the Patient Quality of Life Form encompasses the psychological and social domains of the HRQoL concept, it lacks the physical component. Furthermore all the scores on the 38 items are summed into one overall compound HRQoL score. However, this score is not valid as a single preference value (e.g. utility) as required in economic evaluations.[7]

Some studies attempted to measure and quantify effects on HRQoL using "generic" HRQoL instruments or preference-based HRQoL systems designed for evaluating different kinds of medical interventions. Wyatt et al[8,9] used the Health Utilities Index (HUI, version 2). Harris et al[10] used the CES-D (Center of Epidemiologic Studies-Depression) scale, the SLA (Satisfaction with Life Areas) scale and the QWB (Quality of Well-Being scale) to measure improvement in quality of life and psychological well-being in CI users. The HUI2 and the QWB are suitable to produce a single preference value to arrive at quality-adjusted life years (QALYs) in order to calculate cost-effectiveness ratios. However, these types of instrument cannot be used to evaluate

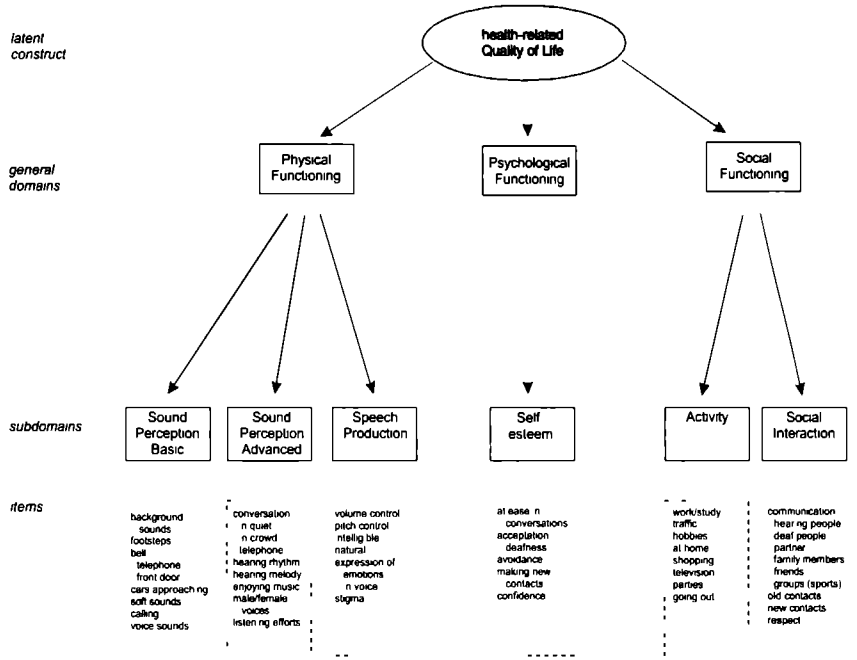

Figure 1. Diagram of the construction of the NCIQ

subjective outcomes and QoL changes induced by CI because they are not sensitive and comprehensive descriptive instruments. The same limitation also applies to the Glasgow Benefit Inventory.[11]

Most existing HRQoL questionnaires for CI users are based on a narrow concept. They focus predominantly on measuring auditory functioning and some associated social activities, so they can hardly be qualified as disease-specific HRQoL instruments. Moreover, most of these questionnaires are based on small sets of items, while other CI-HRQoL questionnaires are not designed to enable adequate statistical testing.

Therefore we decided to develop a standardized questionnaire for CI users that would enable the construction of scores that are reliable, valid and responsive to change. Based on the HRQoL concept, this questionnaire was developed not only to encompass hearing and speech production, but also the psychological and social domains. In the first part of the project we developed a comprehensive and structured CI-HRQoL questionnaire and administered it to CI users and candidates on the waiting list for cochlear implantation.

METHODS

Construction of the HRQoL concept and selection of items.

The development of the Nijmegen Cochlear Implantation Questionnaire (NCIQ) started by formulating the relevant health domains for cochlear implants (CI) users. We followed the conventional approach by measuring health-related quality of life (HRQoL) in which we distinguished three general domains: physical, psychological and social functioning. The following subdomains are specified: sound perception-basic, sound perception-advanced and speech production in the physical domain, while activity and social functioning are specified the social domain. The domain psychological functioning consists of only one subdomain: self-esteem (Figure 1).

Specific features that should be satisfied by the NCIQ were formulated beforehand: 1) each separate subdomain should consist of an equal number of items, 2) all items should be phrased in a similar way, 3) responses to the items should be uniform and 4) the items should be suitable for constructing Likert scales.[12] Additionally, the NCIQ should be a self-report instrument.

The next step consisted of the selection and construction of sets of items for each separate subdomain. Due to the small population of CI users we were unable to follow the conventional psychometric approach for item selection. This empirical psychometric approach is based on: 1) generating a large pool of items, 2) completion of all items by target population, 3) item reduction by the use of psychometric techniques (e.g. factor analysis). Instead of this approach, we have selected items based on intuitive judgement. Numerous items were adapted from other published questionnaires. Several other items were formulated by the authors, based on interviews and previously questionnaires that were used over the years to monitor the rehabilitation of CI users within our team. Each item was formulated as a statement with a 5-point response scale to indicate the degree to which the statement was true. These five response categories were: never (1), sometimes (2), often (3), mostly (4) and always (5) for 55 out of the total of 60 items. The other 5 items were answered according to the CI user's ability to perform the action in question. Response categories for these 5 items were: no (1), poorly (2), moderate (3), adequate (4), good (5). Throughout the questionnaire respondents were also offered a sixth response category to cover items that were not relevant to them.

A pilot version of the questionnaire was sent to five experienced CI users who returned the filled out forms together with their comments to eradicate any questions that were not clear. This information was used to produce the final version of the questionnaire.

Study population and design

The NCIQ was sent together with a letter in which the purpose of the study was explained to 60 adult subjects who consequently received a CI during the period 1989 to

1997 under supervision of the Nijmegen/St. Michielsgestel CI team. All subjects were using oral-aural communication. The selection and implantation procedures have been described previously.[13]

Thirteen of the 60 subjects were excluded from the present study (their results will be presented elsewhere): 10 of these subjects were prelingually deaf and three were postlingually deaf but had been fitted with a single-channel implant. The remaining 47 postlingually deaf adult participants had been fitted with a multichannel implant, using Multi-Peak (MPEAK), Spectral Peak (SPEAK) or Continuous Interleaved Sampling (CIS). They had all been using the implant for at least one year.

The NCIQ was administered twice in a crossover design: once in the past tense to obtain retrospective information and once in the present tense to evaluate the current HRQoL. Half of the CI users filled out the retrospective version first (CI-pre) while the other half filled out the standard version (CI-post). Two weeks after completing and returning the first questionnaire, the other version was sent to the subjects. The retrospective answers of the CI users were compared to the answers from the control group (baseline) of 53 postlingually deaf candidates for CI on the waiting list at our institute.

To study the effect of CI and the construct validity of the NCIQ in more detail, two generic quality of life instruments formed also part of this study together with the assessment by proxies. However, these results will be discussed separately in a forthcoming paper.

Auditory performance

To relate the outcome of the NCIQ to the subjects' auditory performances, we used their results from the Antwerp-Nijmegen (AN) test battery, which is described in detail elsewhere and consists of a number of prerecorded closed-set tests for speech and pattern discrimination. The Spondee Identification Test for the speech discrimination level was used and the Environmental Sounds Identification Test for pattern discrimination. Results obtained one-year post-implant were available in the majority of cases. At this time, most of the subjects will have reached their maximum performance level.

Statistical analysis

Before computation of the six subdomains of the NCIQ, the scores for 27 items of the questionnaire that were phrased in opposite form (see: Appendix), were recoded (6-score). Next, the answer categories (1 ... 5) for all items were transformed: 1=0, 2=25, 3=50, 4=75 and 5=100. Scores for the subdomains were computed by adding together the 10 item scores of each subdomain and dividing by the number of completed items. Missing values and the response category "not applicable" were both treated as not completed. The maximum number of incomplete answers for a specific subdomain was set at three items per subject; above this number the subject was excluded.

Characteristic	CI (n = 45)	Controls (n = 46)
Sex		
Male	46%	60%
Female	54%	40%
Paid employment		
Yes	43%	38%
No	57%	62%
Education level		
Lower	32%	27%
Secondary	50%	57%
Higher	18%	16%
Living situation		
Alone	20%	16%

Table 1. Demographic characteristics of the CI users and the candidates for a CI on the waiting list (control group)

Characteristic	CI (n = 45)	Controls (n = 46)
Age (mean yrs ± SD)	50 ± 16	51 ± 16
Age onset deafness (mean yrs ± SD)	31 ± 18	37 ± 20
Age CI (mean yrs ± SD)	44 ± 16	-
Implant use per day		
0-4 hours	2%	-
4-9 hours	2%	-
9-12 hours	7%	-
12-16 hours	52%	-
more than 16 hours	37%	-
Etiology of deafness		
meningitis	13	5
hereditary	10	10
unknown	6	15
otosclerosis	4	1
trauma	3	2
mumps	1	-
ototoxic drugs	1	3
Widened vestibular aqueduct	2	1
chronic otitis	2	2
Usher's syndrome type II	1	-
Cogan's syndrome	1	-
sudden deafness	1	1
Ostogenesis imperfecta	-	2
Meniere's disease	-	1
Vascular	-	1
Toxicosis	-	1
Cerebellar tumour surgery	-	1

Table 2. Clinical characteristics of the CI users and the candidates for a CI on the waiting list (control group)

For comparison purposes, in both study groups descriptives were computed for the main characteristics: age, age at CI, age at onset deafness, sex, paid employment, education level, living situation and marital status. The internal consistency of the six domains for the NCIQ were assessed by using Cronbach's alpha.[14] An alpha coefficient of 0.70 or higher was considered as sufficient for the purpose of group comparisons.[15] Test-retest reliability was evaluated by readministrating the standard version of the NCIQ to the CI users (n = 43) two months later. Scores for the six domains of the NCIQ were related to the answers on the two audi tory performance tests to evaluate criterion validity. To test the construct validity (relationship 60 items vs. 6 domains) of the NCIQ a confirmatory factor analysis was planned.[16] To assess the responsiveness to change of the NCIQ domains a responsiveness index was estimated by relating change of the CI users (CI post mines CI pre) divided by the variability in stable subjects (test-retest results).[17]

A total of 45 (95%)

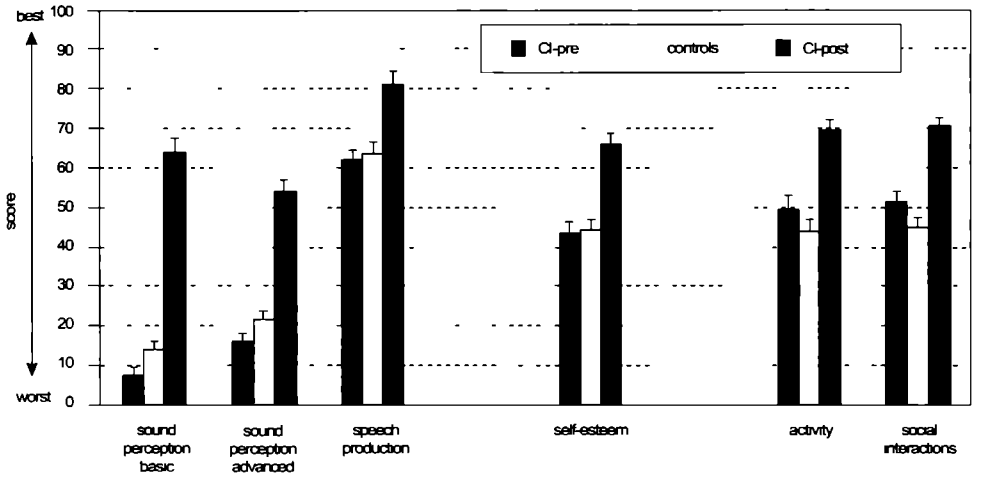

Figure 2. Mean scores (with standard errors of the mean) for the CI users (standard version post, retrospective version pre) and the control group on the six subdomains of the NCIQ.

CI users returned both the standard and the retrospective version of the NCIQ, while 46 controls (87%) completed and returned the standard version of the NCIQ. The main characteristics of the two groups are presented in Table 1 (demographic) and Table 2 (clinical).

Table 1 shows that the two groups were highly comparable. The average daily duration of implant use was more than 12 hours in 89% of the subjects and more than 16 hours in 37% (Table 2). On average, the implant was used for 14 hours per day.

Figure 2 presents the descriptive statistics of the six subdomains for the two groups. Scores from the CI-pre and the controls were similar, whereas the scores from the CI-post assessment were statistically significantly better on all domains (Table 3). The largest differences between CI-post and CI-pre/controls were for sound perception-basic and sound perception-advanced. The differences for the other four subdomains were smaller, although the overall improvement due to a CI was still more than 30% on all four domains. Variation (Table 3: SD) in the answers to the questionnaires was about equal for the six subdomains, for the two groups and for the CI-pre and CI-post assessments. The relatively low standard deviations in the CI-pre and the control assessment for the domains sound perception-basic and sound perception-advanced can be fully attributed to the low mean scores on these two domains.

Table 4 shows the correlation coefficients between the scores on the six domains of the NCIQ and the scores on the Spondee and Environmental Sounds Identification Tests. No obvious correlations were found between the results from these auditory perception tests

Group	Subdomain	mean	S.E.M.	S.D.	range	n
CI users (post)	Sound Perception-Basic	64.1	3.5	23.5	8.3 – 100.0	45
	Sound Perception-Advanced	53.8	2.9	19.6	8.3 – 85.0	45
	Speech Production	81.7	2.7	17.8	21.8 – 100.0	45
	Self-Esteem	66.7	2.5	16.4	10.0 – 95.0	45
	Activity	72.9	2.4	15.9	32.5 – 95.0	45
	Social Interactions	71.9	2.2	14.5	47.2 – 96.9	45
CI users (pre)	Sound Perception-Basic	3.2	0.9	5.8	0.0 – 22.5	45
	Sound Perception-Advanced	14.4	1.7	11.4	0.0 – 50.0	45
	Speech Production	59.8	3.0	20.0	22.5 – 95.0	45
	Self-Esteem	42.0	2.9	19.6	8.3 – 100.0	45
	Activity	49.0	3.1	21.0	7.5 – 97.5	45
	Social Interactions	52.1	2.6	17.2	25.0 – 97.5	45
Control group	Sound Perception-Basic	11.6	2.1	14.4	0.0 – 45.0	46
	Sound Perception-Advanced	19.5	2.0	13.4	2.5 – 69.4	46
	Speech Production	64.6	2.8	18.8	27.5 – 97.5	46
	Self-Esteem	44.8	3.0	20.1	7.5 – 83.3	46
	Activity	45.6	3.4	23.0	8.3 – 87.5	46
	Social Interactions	46.7	2.9	19.8	7.5 – 81.3	46

Table 3. Assessment of the six subdomains of the NCIQ by the CI users (standard version: post, retrospective version: pre) and the control group (standard version)

and the outcomes from the NCIQ.

Table 5 gives internal consistency statistics, test-retest coefficients and responsiveness indices for the six domains. Internal consistency was high (> 0.80) for the two related domains Sound Perception-Basic/Advanced and the domains Activity and Social Interaction. This suggests that those items for these domains are closely related to each other, producing reliable scores for these domains. Moderate Cronbach's alphas (> 0.70) were found for the Speech Production domains and the Self-Esteem domain. Test-retest reliability of the NCIQ proved to be also satisfactory. All domains had coefficients that exceeded 0.60. Unfortunately, due to mathematical restrictions the confirmatory factor analysis to test the construct validity of the NCIQ could not be carried out. The responsiveness indices of the six domains are also presented in Table 5. All six domains appeared to be very sensitive for measuring changes resulting from cochlear implantation. The responsiveness indices were all greater than 1.

DISCUSSION

The purpose of this study was to develop a quantifiable, self-assessment health-related

Subdomain	Spondee Identification Test	Environmental Sounds Identification Test
Sound Perception-Basic	0.23	0.38
Sound Perception-Advanced	0.49	0.54
Speech Production	0.36	0.59
Self-Esteem	0.38	0.23
Activity	0.40	0.43
Social Interactions	0.32	0.36

Table 4. Correlations of the six subdomains of the NCIQ from the CI users (n = 45) with the Spondee Identification Test and the Environmental Sounds Identification Test

quality of life (HRQoL) instrument for use in Cochlear Implant (CI) users. This questionnaire (the NCIQ) was then administered to an existing CI population and their answers were compared to those from deaf controls on the waiting list for a CI. In this way we obtained valuable information about the HRQoL of users and potential users. So far, no comprehensive "disease"-specific HRQoL instrument has been developed for CI users, although a CI can be expected to have a considerable impact on the quality of life in this group.

The benefits of cochlear implantation were illustrated by the fact that most of the subjects (89%) were using their CI for more than 12 hours a day. This is in agreement with the findings of Kelsall et al (18) and Summerfield and Marshall (1): 81% and 85% were using the implant for more than 10 hours a day 9 months after the operation respectively. The average duration of implant use per day in our population was 14 hours, which is in agreement with the literature.

The NCIQ measured important improvements in all six domains between the CI-pre and CI-post assessments. Great improvement in the perceived quality of life was not only measured for the Physical domains, but also for the Psychological and Social domains. The largest improvement was recorded for the sound perception-basic subdomain and sound perception-advanced subdomain, which is understandable because deaf subjects score next to nothing on these domains. Improvements were found for all the items in these two subdomains, even for listening to music, despite the fact that the processors of the implant systems are specially designed for processing speech, not music. We feel that being able to enjoy music can contribute considerably to a sub-

Subdomain	Internal consistency (n = 45)	Test-retest reliability (n = 35)	Responsiveness index
Sound Perception-Basic	0.81	0.83	4.59
Sound Perception-Advanced	0.84	0.85	3.93
Speech Production	0.73	0.78	1.81
Self-Esteem	0.75	0.64	1.68
Activity	0.89	0.82	2.36
Social Interactions	0.84	0.81	2.09

Table 5. Internal consistency (Cronbach's alpha), test-retest reliability (intraclass correlation coefficient) and responsiveness index of the assessment of the six subdomains of the NCIQ by the CI users

ject's quality of life; consequently, music should not be categorized at the same level as environmental sounds, but rather on a more cognitive level as has been suggested by Gfeller et al.[19] Therefore we integrated music items with those concerning speech discrimination in the sound perception-advanced subdomain. For the other domains there was a certain preoperative baseline. However, the benefits of a CI were also reflected on these domains by an average rise in score of 30%.

In this study we also examined the psychometric characteristics of the NCIQ. In advance of the discussion of the NCIQ results it is important to be noted that we observed a strong agreement between the retrospective answers of the CI users regarding their pre-implant HRQoL and the HRQoL presently perceived by the deaf candidates on the waiting list for CI. This strong resemblance between the CI users and the controls in this study provides support for the validity of interpreting the retrospective information.

There did not appear to be any prominent positive correlation between the HRQoL scores of the six NCIQ domains and objective measures of performance as measured by the two auditory perception tests (Table 4). Especially on the sound perception domains we expected higher correlations, which would confirm the criterion validity for at least two domains of the NCIQ.

An explanation for this low association might be the small range in outcomes on the sound perception tests which inflate the differentiation among the scores of the subjects. On the other hand it can be argued that the patients subjective perception of benefits due to CI is not directly linked to the objective performance levels. From an earlier study (20), we know that despite their modest auditory perception, prelingually deaf adults also keep using their implants and report substantial benefits regarding their quality of life. The effect that a CI has on the quality of life of these patients might even outweigh any improvements in hearing ability measured by conventional speech perception tests. This emphasizes the need for a comprehensive and structured CI-HRQoL instrument in addition to existing sound and speech perception tests to evaluate the outcome of cochlear implantation. However, others found modest positive correlations between sound perception performance and overall quality of life,[1,6] but the populations included prelingually deaf subjects and subjects with a single-channel system. The large range of performance levels measured might explain these correlations.

For four of the six domains the NCIQ exhibited high levels of internal consistency. Lower, though generally acceptable reliability estimates were found for the domains Speech Production and Self-Esteem. Test-retest results for the NCIQ were satisfactory. An important concern in the development of any measurement instrument is to develop an instrument that is both reliable and valid. This last aspect could not be formally examined in this study in detail. In particular the construct validity of the NCIQ could not be studied due to the small number of observations in relation to the large number of questions. However, in a forthcoming publication we will focus on comparing the results

of the NCIQ to those from other more generic quality of life instruments to further evaluate the construct/criterion validity of this instrument. Finally, the NCIQ proved to be a sensitive instrument for detecting change in deaf adults due to a CI on all six domains.

Despite the fact that additional research is required, we think that the NCIQ has considerable potential for evaluating the effect of CI in adults. Its good internal consistency, its content validity (Figure 1), its probable construct validity, its good responsiveness and its ease of administration and scoring all suggest that is a useful instrument for the evaluation of the outcomes of cochlear implantation.

Standardization in this type of research is highly recommended, because it will enable comparisons between different CI populations or studies and create the opportunity to monitor the effect of technical improvements to CIs in the future. Such core "disease"-specific HRQoL questionnaires have already been developed and used in several other areas, for instance, cancer and chronic rheumatic disorders. The results of this study highlight the impressive merits of cochlear implantation that seem comparable or even greater than numerous other medical treatments for (non-lethal) diseases.

REFERENCES

1 Summerfield, Q A and Marshall, D A Cochlear Implantation in the UK 1990-1994 London HMSO 1995

2 Blamey P, Arndt P, Bergeron F, Bredberg G, Brimacombe J, Facer G, Larky J, Lindstrom B, Nedzelski J, Peterson A, Shipp D, Shaller S, Whitford L Factors affecting auditory performance of postlinguistically deaf adults using cochlear implants Audio Neurootol 1996,1 293-306

3 East CA and Cooper HR Extra-cochlear implants the patient's viewpoint British Journal of Audiol 1986,2055-9

4 Tyler RS and Kelsay D Advantages and disadvantages reported by some of the better cochlear implant patients Am J Otol 1990,11282-9

5 Zhao F, Stephens SDG, Sim SW, Meredith R The use of qualitative questionnaires in patients having and being considered for cochlear implants Clin Otolaryngol 1997,22 254-9

6 Maillet CJ, Tyler RS, Jordan HN Change in the quality of life of adult cochlear implant patients Ann Otol Rhinol Laryngol 1995,165(suppl) 31-48

7 Gold MR, Siegel JE, Russel LB, et al Cost-Effectiveness in Health and Medicine New York Oxford University Press, 1996

8 Wyatt JR, Niparko JK, Rothman ML, deLissovoy G Cost effectiveness of the multichannel cochlear implant Am J Otol 1995,16(1) 52-62

9 Wyatt JR, Niparko JK, Rothman ML Cost utility of the multichannel cochlear implant in 258 profoundly deaf individuals Laryngoscope 1996,106 816-21

10 Harris JP, Anderson JP, Novak R An outcomes study of cochlear implants in deaf patients Audiologic, economic and quality of life changes Arch Otolaryngol Head Neck Surg 1995,121 398-404

11 Robinson K, Gatehouse S, Browning GB Measuring patient benefit from otolaryngological surgery and therapy Ann Otol Rhinol Laryngol 1996,105 415-22

12 Streiner DL, Norman GR Health Measurement Scales a practical guide to their development and use Oxford Oxford University Press, 1995

13 Broek van den P, Admiraal R Selection of patients for a cochlear implant Acta Otorhinolaryngologica Belg 1991,45 243-6

14 Cronbach LJ Coefficient alpha and the internal structure of tests Psychometrika 1951,16(3) 297-334

15 Essink-Bot ML, Krabbe PFM, Bonsel GJ, et al An empirical comparison of four generic health status measures the Nottingham Health Profile, the MOS 36-Item Short-Form Health Survey, the COOP/WONCA charts, and the EuroQol instrument Medical Care 1997,35(5) 522-37

16 Joreskog KG, Sorbom D Advances in Factor Analysis and Structural Equation Models Cambridge, Massachusetts Abt Books, 1979

17 Deyo RA, Diehr P, Patrick DL Reproducibility and responsiveness of health status measures statistics and strategies for evaluation Controlled Clinical Trials 1991,12 142S-58S

18 Kelsall DC, Shallop JK, Burnelli T Cochlear implantation in the elderly Am J Otol 1995,16(5) 609-15

19 Gfeller K, Knutson JF, Woodworth G, et al Timbral recognition and appraisal by adult cochlear implant users and normal-hearing adults J Am Acad Audiol 1998,9 1-19

20. Hinderink JB, Mens LHM, Brokx JPL, et al. Performance of prelingually and postlingually deaf patients using single-channel or multichannel cochlear implants. Laryngoscope 1995;105:618-22.

The Nijmegen Cochlear Implant Questionnaire (NCIQ)

Please answer the following 60 questions regarding the CI situation (only if none of the possibilities is applicable use "not applicable")

	never	sometimes	regularly	usually	always	N/A
1. Can you hear background noises (toilet flushing, vacuum cleaner)?	☐	☐	☐	☐	☐	☐
2. Does your hearing impairment form a serious obstacle in your contact with people with normal hearing?	☐	☐	☐	☐	☐	☐
3. Are you able to whisper if you have to?	☐	☐	☐	☐	☐	☐
4 Do you feel at ease in company despite your hearing impairment?	☐	☐	☐	☐	☐	☐
5 Can you hold a conversation in a quiet environment (with or without lip-reading) with one person?	☐	☐	☐	☐	☐	☐
6 Does your hearing impairment form a serious problem during your work or studies?	☐	☐	☐	☐	☐	☐
7 Can you hear the footsteps of other people in your the house (e g in the hall or on the stairs)?	☐	☐	☐	☐	☐	☐
8. Does your hearing impairment form a serious problem in your contact with deaf people?	☐	☐	☐	☐	☐	☐
9 Are you able to shout if you need to?	☐	☐	☐	☐	☐	☐
10. Does it bother you that you are hard of hearing?	☐	☐	☐	☐	☐	☐
11. Are you able to hold a conversation with two or more persons in a quiet environment (with or without lip-reading)?	☐	☐	☐	☐	☐	☐
12. Does your hearing impairment form a serious problem in traffic?	☐	☐	☐	☐	☐	☐
13 Can you hear your own telephone or doorbell ringing?	☐	☐	☐	☐	☐	☐
14 Does your hearing impairment form a serious problem when you are with a group of people (hobbies, sport, holidays)?	☐	☐	☐	☐	☐	☐
15 Are you able to make yourself understood to strangers without using hand gestures?	☐	☐	☐	☐	☐	☐
16. Do you become irritated if you cannot follow a conversation?	☐	☐	☐	☐	☐	☐
17. When you are in a busy shop, can you understand the shop assistant?	☐	☐	☐	☐	☐	☐
18. Does your hearing impairment form a serious problem during leisure-time activities?	☐	☐	☐	☐	☐	☐

		never	sometimes	regularly	usually	always	N/A
19	Can you hear (not feel) the front door slam when you are busy at home?	☐	☐	☐	☐	☐	☐
20	Does your hearing impairment form a serious problem in your contact with the people you live with (family/partner)?	☐	☐	☐	☐	☐	☐
21	Are you able to adapt your voice to different situations (noisy environment, quiet environment)?	☐	☐	☐	☐	☐	☐
22	Do you avoid speaking to strangers?	☐	☐	☐	☐	☐	☐
23	Are you able to enjoy music?	☐	☐	☐	☐	☐	☐
24	Does your hearing impairment form a serious problem for functioning in the home?	☐	☐	☐	☐	☐	☐
25	Are you able to hear cars approaching in traffic?	☐	☐	☐	☐	☐	☐
26	Are you left aside in company because of your hearing impairment?	☐	☐	☐	☐	☐	☐
27	Can strangers hear from your voice that you are deaf or hearing impaired?	☐	☐	☐	☐	☐	☐
28	Do you ask other people to speak more loudly or clearly if they are speaking too softly or unclearly?	☐	☐	☐	☐	☐	☐
29	Are you able to recognise certain melodies in music?	☐	☐	☐	☐	☐	☐
30	Does your hearing impairment form a serious problem when your are shopping?	☐	☐	☐	☐	☐	☐
31	Can you hear soft noises (key falling, microwave beeping)?	☐	☐	☐	☐	☐	☐
32	Do you go places where your hearing impairment might form a serious handicap?	☐	☐	☐	☐	☐	☐
33	Can you make yourself understood to acquaintances without using hand gestures?	☐	☐	☐	☐	☐	☐
34	Do you feel anxious when talking to strangers?	☐	☐	☐	☐	☐	☐
35	Are you able to recognise certain rhythms in music?	☐	☐	☐	☐	☐	☐
36	Does your hearing impairment form a serious problem when watching television?	☐	☐	☐	☐	☐	☐
37	Can you hear (not feel) someone approaching you from behind?	☐	☐	☐	☐	☐	☐
38	Does your hearing impairment form a serious hindrance in your contact with people who live in your neighbourhood?	☐	☐	☐	☐	☐	☐
39	How often does it annoy you that people can hear from your voice/speech that you have a hearing problem?	☐	☐	☐	☐	☐	☐
40	Can you understand strangers without lip-reading?	☐	☐	☐	☐	☐	☐

	never	sometimes	regularly	usually	always	N/A
41 Does your hearing impairment form a serious problem at parties (e g birthday)?	☐	☐	☐	☐	☐	☐
42 Can you hear it (not necessarily understand it) when a person starts talking on the radio?	☐	☐	☐	☐	☐	☐
43 Does your hearing impairment form a serious problem when you are with friends?	☐	☐	☐	☐	☐	☐
44. Can you make contact easily with other people despite your hearing problem?	☐	☐	☐	☐	☐	☐
45. Can you hear the difference between a man's voice, a woman's voice, and a child's voice?	☐	☐	☐	☐	☐	☐
46 Does your hearing impairment forms a serious problem when dealing with formal matters (insurance, solicitor, municipal office)?	☐	☐	☐	☐	☐	☐
47 Can you hear it when someone calls you?	☐	☐	☐	☐	☐	☐
48 Does your hearing impairment form a serious problem in your contacts with family members?	☐	☐	☐	☐	☐	☐
49 Are there situations in which you would feel happier if you were not hearing-impaired?	☐	☐	☐	☐	☐	☐
50 Do you feel it tiring to listen (with or without lip-reading)?	☐	☐	☐	☐	☐	☐
51. Does your hearing impairment form a serious problem when you go out or go on trips?	☐	☐	☐	☐	☐	☐
52. Can you hear voices from another room (e g children playing, baby crying)?	☐	☐	☐	☐	☐	☐
53 When you are in a group, do you feel that your hearing impairment keeps people from taking you seriously?	☐	☐	☐	☐	☐	☐
54 Does your hearing impairment undermine your self-confidence?	☐	☐	☐	☐	☐	☐
55 Does your hearing impairment prevent you from sticking up for yourself (at work, in relationships)?	☐	☐	☐	☐	☐	☐

Please note the answer categories for the following 5 questions are changed

	no	poor	fair	good	quite well	N/A
56 Are you able to make your voice sound angry, friendly or sad?	☐	☐	☐	☐	☐	☐
57. Can you control the pitch of your voice (high, low)?	☐	☐	☐	☐	☐	☐
58 Can you control the volume of your voice?	☐	☐	☐	☐	☐	☐
59. Can you make your voice sound 'natural' (so that is does not sound like a deaf person's voice)?	☐	☐	☐	☐	☐	☐
60. Are you able to hold a simple telephone conversation?	☐	☐	☐	☐	☐	☐

Code book

Domain	Question	Recoding (6-score)
Physical		
Sound Perception-Basic	1, 7, 13, 19, 25, 31, 37, 42, 47, 52	
Sound Perception-Advanced	3, 9, 15, 21, 27, 33, 56, 57, 58, 59	27
Speech Production	5, 11, 17, 23, 29, 35, 40, 45, 50, 60	50
Psychological		
Self-Esteem	4, 10, 16, 22, 28, 34, 39, 44, 49, 54	10, 16, 22, 34, 39, 49, 54
Social		
Activity Limitations	6, 12, 18, 24, 30, 36, 41, 46, 51, 55	6, 12, 18, 24, 30, 36, 41, 46, 51, 55
Social Interactions	2, 8, 14, 20, 26, 32, 38, 43, 48, 53	2, 8, 14, 20, 26, 38, 43, 48

CHAPTER SEVEN

THE EFFECT OF COCHLEAR IMPLANT USE IN POSTLINGUALLY DEAF ADULTS

Paul F.M. Krabbe, PhD; Johannes B. Hinderink, MD;
Paul van den Broek, MD, PhD

International Journal of Technology Assessment in Health Care 16: 2000 pp. 864-873

THE EFFECT OF COCHLEAR IMPLANT USE IN POSTLINGUALLY DEAF ADULTS

Paul F.M. Krabbe, PhD; Johannes B. Hinderink, MD;
Paul van den Broek, MD, PhD

ABSTRACT

Objective: To assess the effect of the use of cochlear implants (CI) on the health status of postlingually deaf adults.
Methods: Participants comprised 45 postlingually deaf adult multichannel CI users and 46 deaf candidates on the waiting list for a CI. The latter group acted as controls to corroborate the validity of retrospective completion of the questionnaires by the CI recipients. Three HRQoL instruments were used: 1) a specially developed CI questionnaire (NCIQ), 2) a generic HRQoL questionnaire (SF-36) and 3) a health-state classification system (HUI-2) suited to estimate single preference scores.
Results: Retrospectively estimated pre-implant scores in the CI user group corresponded very well with the scores in the control group. Post-implant scores in the CI users were substantially higher in all six domains ($p < 0.001$) of the NCIQ than the scores in the controls. Effects due to a CI were also observed with the SF-36 in five out of the seven domains ($p < 0.01$). Statistically significant differences between the two groups ($p = 0.001$) were observed in two of the six domains of the HUI-2.
Conclusions: All three questionnaires detected improvements in HRQoL due to CI use. To make a detailed assessment of the effect of a CI on functional outcomes and well-being a special-purpose HRQoL instrument is far more adequate than a general HRQoL instrument. This study also showed that a CI affects several other health domains besides auditory performance. The effect of CI use on general functioning and well-being proved to be considerable.

INTRODUCTION

Cochlear implantation in profoundly deaf people is a relatively new medical technology that partially restores auditory perception. The obvious benefits of this technique are enhancement of sound and speech perception and speech production. Owing to the technological development of cochlear implants (CI), the results of this medical intervention have improved over the past few decades.[1,2] Besides the re-establishment of sound perception and the resulting improvement in speech production, the use of a CI may also have a positive impact on other health domains.

Clinicians and policy makers have recognized that changes in a patient's health status are the primary outcome of medical interventions. Subsequently, the WHO has extended the definition of health with psychological and social domains. More and more medical interventions are being evaluated in a comprehensive manner, by looking at a broad range of health domains that can be affected by deterioration in a person's general health status. The paradigm that is focused on the comprehensive measurement of health outcomes on the subjective level of the patient is referred to as "health-related quality of life" (HRQoL). In the case of a CI, this means that evaluations should also extend to general aspects such as: self-esteem, daily activities and social functioning.

So far, most of the reports on HRQoL aspects in CI users have generally been based on the use of open-ended questionnaires or on interviews with patients.[3-7] Only one study on the impact of CI use made use of closed-set questionnaires, which were built up in a systematic manner and provided quantifiable scores.[8] Although this questionnaire encompassed the psychological and social domains of the HRQoL concept, it lacked the physical component. In addition, the psychological and social domains were not dealt with separately, but were aggregated into one composite score.

Some studies attempted to measure and quantify HRQoL effects in CI users with "generic" HRQoL instruments or preference-based HRQoL systems designed to evaluate different types of medical intervention. Wyatt et al.[9,10] used the Health Utilities Index (HUI, version 2). Harris et al.[11] used the Quality of Well-Being scale (QWB) to measure improvement in quality of life and psychological well-being in CI users. The HUI-2 and QWB were specifically developed to produce a single preference score or value (technical term: utility), in order to merge HRQoL values with survival data to calculate quality-adjusted life years (QALYs). However, these types of instrument cannot be used to make detailed subjective evaluation of outcomes or HRQoL changes induced by CI use, because they are not sensitive or comprehensive enough.

To assess the effect of CI use, we have used three conceptually different HRQoL questionnaires. The first instrument can be classified as a "disease"-specific HRQoL questionnaire for self-report use, specifically developed for the evaluation of CI user

populations. In addition, two generic HRQoL questionnaires were used that are not restricted solely to the evaluation of CI user populations and can therefore be used to compare the effect of CI use on HRQoL to the results of other medical interventions. The primary aim of this study was to assess the effect of CI use on the perceived health status of adults with profound sensorineural hearing loss.

METHODS

Quality of Life Measures

The Nijmegen Cochlear Implantation Questionnaire (NCIQ) is a disease-specific instrument based on the conventional approach to measuring health-related quality of life (HRQoL).[12] Three general domains are distinguished: physical, psychological and social functioning. The following subdomains are specified: sound perception-basic, sound perception-advanced and speech production in the physical domain, while activity and social functioning are specified in the social domain. The domain psychological functioning consisted of only one subdomain: self-esteem. Each domain consists of 10 items, formulated as a statement with a 5-point response scale to indicate the degree to which the statement applies to the respondent. There is also a sixth response category if the item is not considered relevant. Scores range from 0 to 100 (optimal).

The Medical Outcome Study Short-Form 36 (SF-36) was developed in the United States on the basis of the large battery of health status and HRQoL instruments employed in the Medical Outcomes Study (13). This generic, e.g., non-disease-specific, HRQoL questionnaire consists of 36 items. These items are organized into eight domains: physical functioning (10 items), social functioning (2), problems with role functioning due to physical problems (4), problems with role functioning due to emotional problems (3), pain (2), mental health (5), vitality (4) and an overall domain general health perception (5). The number of response choices per item ranges from two to six. The SF-36 yields an 8-dimensional profile, in which each domain has a range from 0 to 100 (optimal). The Dutch version of the SF-36 employed in the current study was developed as part of the International Quality of Life Assessment (IQOLA) Project.[14] Psychometric properties of this instrument have been studied in detail and are considerable adequate.[15-18]

At present the Health Utility Index, Mark II (HUI-2) is probably the most comprehensive multi-attribute health-state classification system.[19] It is focused on the more functional concepts of HRQoL, such as disabilities (dysfunction) and resulting dependencies. The HUI-2 encompasses seven domains (sensation, mobility, emotion, cognition, self-care, pain and fertility). Obviously, the last domain can safely be omitted if it is not relevant. Three to five levels of functioning are defined in each domain (also called "attributes"). Any specific combination of the applicable number of domain levels

constitutes a unique health state. Values for the attributes range from 0 to 100 (optimal). A distinguishing feature of the HUI-2 – as opposed to for example the SF-36 and the NCIQ – is the potential to assign a numerical value (or utility) to any health status of a particular participant based on the HUI-2 classification. Each attribute has an associated weight that indicates the subjective assessment of the attribute in question. This utility on a scale from 0 to 1.0 (0 = death, 1.0 = perfect health) is obtained by applying a predetermined multi-attribute utility function. Utility data express the overall assessment of a specific health status and these can be merged together with expected life years to compute Quality-Adjusted Life Years (QALYs). QALYs are appealing, because they provide a relatively simple means of reflecting the HRQoL effects of medical interventions and enable comparisons with interventions that have very different types of outcome (e.g., in cost-effectiveness analysis).

Study Population and Design

In April 1998 the NCIQ was sent together to 60 adult subjects who had received a CI during the period 1989 to 1997 under the supervision of the Nijmegen/St. Michielsgestel CI team with a letter explaining the purpose of the study. All the subjects were using oral-aural communication. The selection and implantation procedures have been described previously.[20]

Thirteen out of the 60 subjects were excluded from the study: 10 of these subjects were prelingually deaf and three were postlingually deaf but had been fitted with a single-channel implant. The remaining 47 postlingually deaf adult participants have been fitted with a multichannel implant, using advanced speech encoding strategies (MPEAK, SPEAK, CIS). They have been using their implant for at least one year.

The three questionnaires were administered twice to the CI users in a crossover design: once in the past tense to obtain retrospective information and once in the present tense to evaluate the current HRQoL. Half of the CI users filled out the retrospective version first (CI-pre), while the other half filled out the standard (present tense) version (CI-post). Two weeks after completing and returning the first first, the other version was sent to the CI users. Results from both versions are presented. The retrospective answers of the CI users were compared to those from the control group (baseline) of postlingually deaf candidates on the waiting list for a CI at our institute.

Statistical Analysis

Descriptive statistics of the two study groups were computed for the main characteristics: age, age at cochlear implantation, age at onset deafness, sex, paid employement, education and living situation. Scores on the three questionnaires were declared as missing values if nothing was filled in or if ambiguous information was

provided. On the NCIQ missing values and the response category "not applicable" were both treated as not completed. The maximum permitted number of incomplete answers for a specific subdomain was set at three items per subject; above this number the subject was excluded.

As the distribution of the scores on the majority of separate domains was highly skewed (as evidenced by the results of Kolmogorov-Smirnov tests), nonparametric tests were used to analyse whether the scores of the two groups were significantly different. Wilcoxon's Signed Rank test was used to compare the dependent scores of the CI group. The Mann-Whitney U test was applied to test the two assessments of the CI group against the response of the controls. To avoid the effect of multiple testing, $p<0.01$ was regarded as statistically significant. As the sample size largely determined whether an effect would be statistically significant, we employed an estimator of effect size d for continuous variables.[21] Effect sizes were calculated by dividing the means of the two measures by the deviation in that scale.

Characteristic	CI (n = 45)	Controls (n = 46)
Sex		
Male	46%	60%
Female	54%	40%
Paid employment		
Yes	43%	38%
No	57%	62%
Education level		
Lower	32%	27%
Secondary	50%	57%
Higher	18%	16%
Living situation		
Alone	20%	16%
With others (partner, children)	80%	82%
Care centre	0%	2%
Age (mean yrs ± SD)	50 ± 16	51 ± 16
Age onset deafness (mean yrs ± SD)	31 ± 18	37 ± 20
Duration deafness (yrs ± SD)	13 ± 12	16 ± 14
Age CI (mean yrs ± SD)	44 ± 16	-
CI use (yrs ± SD)	5 ± 2.8	-

Table 1. Demographic and clinical characteristics of the cochlear implant (CI) users and the candidates for a CI on the waiting list (control group)

RESULTS

Respondent Characteristics

A total of 45 (95%) CI users returned both the standard and the retrospective version of the NCIQ, the SF-36 and the HUI-2. In the control group, 46 subjects (87%) completed the standard and the adapted (CI-post) version of the three questionnaires. Demographic and clinical characteristics of the two groups were very similar (Table 1).

Domain	Control Group (n = 46)	CI Group (n = 45)		CI post - CI pre (n = 45)		
		CI pre	CI post	Change (S.D.)	*p* Value*	Effect size**
Sound perception-basic	11.6 (14.4)	3.2 (5.8)	64.1 (23.5)	61.3 (23.9)	< 0.001	3.56
Sound perception-advanced	19.6 (13.4)	14.4 (11.4)	53.8 (19.6)	40.5 (16.7)	< 0.001	2.46
Speech production	64.6 (18.8)	59.8 (20.1)	81.7 (17.8)	22.2 (21.2)	< 0.001	1.15
Self-esteem	44.8 (20.1)	42.0 (19.6)	66.7 (16.4)	25.6 (17.3)	< 0.001	1.37
Activity	45.6 (23.0)	49.0 (21.0)	72.9 (15.9)	24.7 (15.6)	< 0.001	1.28
Social interactions	46.7 (19.8)	52.1 (17.2)	71.9 (14.5)	20.1 (11.0)	< 0.001	1.24

*Nonparametric testing: Wilcoxon Signed Ranks Test (2-tailed Monte Carlo sampling).
** Effect size: $d \leq 0.2$ indicates a small effect, $d \cong 0.5$ a medium effect and $d \geq 0.8$ a large effect.

Table 2. NCIQ: Mean scores (S.D. between parentheses) on the six domains of the Nijmegen Cochlear Implant Questionnaire (NCIQ) of the cochlear implant (CI) users (retrospective version = CI pre, standard version = CI post) and the control group (baseline measurement)

NCIQ

Table 2 shows the mean scores on the six domains of the NCIQ together with their standard deviations. On the NCIQ, scores were substantially higher on all six domains during CI use. Differences between the CI-pre and CI-post scores were all statistically significant ($p < 0.001$). The largest difference between CI-post and CI-pre (and the controls) was observed for the domains sound perception-basic and sound perception-advanced. Differences were smaller for all other four domains although the overall improvement owing to the CI was still more than 30%. Moreover, the effect sizes were large for all six domains ($d > 0.8$). CI-pre and control scores were very similar, expect for sound perception-basic.

SF-36

Higher scores for the CI-post period were also observed on three domains of the SF-36 (Table 3). These domains were: social functioning, the two role functioning domains (physical and emotional) and the mental health domain. Except for the domains pain and vitality, all the differences between the CI-pre and CI-post scores were statistically significant ($p \# 0.01$). The effect of a CI on physical functioning was negative and the effect size was small ($d = 0.27$). CI-pre and control scores on the SF-36 were fairly similar, but the differences between the two groups were greater than they were for the NCIQ. There was no systematic effect between the control group and the CI-pre assessments.

Domain	Control Group	CI Group (n = 45)		CI post CI pre (n = 45)		
	(n = 46)	CI pre	CI post	Change (S D)	*p* Value*	Effect size**
Physical functioning	79 2 (24 8)	89 7 (17 1)	84 5 (21 5)	3 7 (10 5)	0 01	0 27
Social functioning	72 8 (29 6)	58 6 (27 4)	84 7 (20 2)	26 5 (27 3)	< 0 001	1 08
Role functioning (Physical)	60 2 (41 5)	60 6 (40 1)	80 0 (35 6)	19 9 (40 2)	0 002	0 51
Role functioning (Emotional)	72 7 (35 6)	64 4 (38 3)	85 2 (32 2)	20 9 (40 5)	0 003	0 59
Pain	77 2 (25 8)	88 7 (17 3)	83 2 (17 1)	5 3 (17 8)	N S	0 32
Mental Health	71 0 (21 0)	61 6 (18 9)	77 3 (17 9)	15 8 (19 9)	< 0 001	0 85
Vitality	66 4 (20 2)	67 9 (18 6)	71 5 (18 7)	3 4 (18 5)	N S	0 19
General health perception	68 7 (21 5)	***	72 3 (19 8)			

*Nonparametric testing Wilcoxon Signed Ranks Test (2 tailed Monte Carlo sampling)
** Effect size d ≤ 0 2 indicates a small effect, d ≅ 0 5 a medium effect and d ≥ 0 8 a large effect
***Not measured

Table 3 SF-36 Mean scores (S D between parentheses) on the eight domains of the MOS Short Form 36 (SF 36) instrument of the cochlear implant (CI) users (retrospective version = CI pre, standard version = CI post) and the control group (baseline measurement)

HUI-2

The HUI-2 domains (Table 4) showed less significant results than those of the other two HRQoL questionnaires Only two out of the six domains showed statistically

Domain	Control Group	CI Group (n = 45)		CI post - CI pre (n = 45)		
	(n = 46)	CI pre	CI post	Change (S D)	*p* Value*	Effect size**
Sensation	14 5 (18 2)	3 0 (9 6)	48 8 (23 1)	46 4 (24 2)	< 0 001	2 59
Mobility	92 4 (17 4)	97 8 (9 0)	97 8 (9 0)	0 0 (5 4)	N S	0
Emotion	89 1 (13 6)	79 5 (19 6)	90 6 (13 4)	10 5 (17 5)	0 001	0 66
Cognition	83 8 (19 8)	93 2 (13 8)	95 5 (13 7)	1 6 (12 5)	N S	0 17
Self Care	97 1 (11 9)	99 2 (5 1)	98 5 (7 1)	-0 7 (5 1)	N S	0 11
Pain	88 6 (18 8)	90 3 (13 4)	88 3 (16 5)	1 7 (15 8)	N S	0 13
HUI 2 utility	0 62 (0 16)	0 55 (0 11)	0 82 (0 14)	0 28 (0 15)	< 0 001	2 08

*Nonparametric testing Wilcoxon Signed Ranks Test (2 tailed Monte Carlo sampling)
** Effect size d ≤ 0 2 indicates a small effect, d ≅ 0 5 a medium effect and d ≥ 0 8 a large effect

Table 4 Mean scores (S D between parentheses) on the six domains of the Health Utility Index (HUI version 2)of the cochlear implant (CI) users (retrospective version = CI pre, standard version = CI post) and the control group (baseline measurement)

Domain	Norm scores General population (n = 2,474)*	CI CI study group (n=45)			Renal replacement therapy*			Heart transplantation**		
		Postlingually deaf adults (Pre)	Adult CI users (Post)	Mean change (Post - Pre)	Haemo-dialysis (n=43)	RTx (n=102)	Mean change (Rtx - Haem.)	Waiting list (n=42)	HTx (n=143)	Mean change (Htx – wait. L.)
Physical functioning	84	90	85	-5	46	68	22	36	71	35
Social functioning	83	59	85	26	54	80	26	63	85	22
Role functioning (Physical)	81	61	80	19	51	63	12	27	62	35
Role functioning (Emotional)	81	64	85	21	75	80	5	71	77	6
Pain	75	89	83	-6	82	78	-4	60	69	-9
Mental Health	75	62	77	15	66	79	13	75	77	2
Vitality	61	68	72	4	41	63	22	39	62	23
General health perception	72	-***	72	-	42	64	22	33	70	37

* Data from Khan et al, 1995
** Data from Rector and Kubo, 1993
*** Not measured

Table 5: CI versus some other medical interventions
Mean scores (S.D. between parentheses) of the cochlear implant (CI) study group (retrospective version = pre, standard version = post) on the domains of the MOS Short-Form 36 (SF-36) instrument, norm scores of the SF-36 and scores obtained with the SF-36 for other diseases

significant differences between the CI-pre and CI-post scores: sensation (partially comprising hearing functioning) and emotion. The effect size observed for sensation was large ($d > 0.8$). Utilities obtained with the HUI-2 increased from 0.55 (CI-pre) to 0.82 (CI-post), which is a large effect ($d = 2.08$).

Generally, the standard deviations for the twenty domains of the three HRQoL questionnaires were moderate. Relatively large standard deviations were observed for two SF-36 domains (role functioning domains, whereas the smallest standard deviations were observed for the HUI-2 weights.

DISCUSSION

Our study showed that a cochlear implant (CI) led to a significant improvement in health-related quality of life (HRQoL) of post-lingually deaf adults. We used three HRQoL questionnaires, each of them based on a different HRQoL measurement approach. The disease-specific Nijmegen Cochlear Implantation Questionnaire (NCIQ) measured important improvements on six health domains between the CI-pre and CI-post situation. As expected, major improvements were observed on the three domains focused

on sound and speech functionality Moreover, an average increase in scores of 30% was observed on the psychological domain and the two social domains The generic Medical Outcome Short-Form 36 (SF-36) questionnaire reflected significant effects on four of its seven domains social functioning, role functioning (physical and emotional) and mental health The more crude classification system of the Health Utility Index (HUI-2) showed great improvement in sensation (comprising vision, speech and hearing) and a slightly smaller improvement in emotion Standard deviations of the HUI-2 classification were smaller than those of the two questionnaires, which is basically an intrinsic feature of any concise instrument

There was a strong agreement between the retrospective answers of the CI users regarding their pre-implant HRQoL and the HRQoL perceived by the deaf candidates on the waiting list for a CI This agreement between the CI users and the controls provides strong support for the validity of interpreting retrospective information from CI users Differences between the CI-pre scores and the scores of the controls may be attributed to different aspects It is well-known that valuation of health states may differ according to illness experience [22,23] People with a disability sometimes manage to adapt in such a way, that their HRQoL assessments of their own health status even exceed those of healthy controls This phenomenon has been the subject of study in the social sciences under the heading of cognitive dissonance and valuation- or response-shift [24] A similar process may also be applicable to the responses of the non-CI users (controls) If the differences between the CI-pre scores and the scores of the control group were largely thought to originate from adaptation effects, than we may consider the CI-pre scores in this retrospective study as more precise HRQoL estimates than the scores of the controls Another possible explanation is that the retrospective answers to the CI questionnaires may have been confounded by inaccurate memory, although this would probably only have led to an increase of the unreliability of the scores but not to systematically biased responses [25] Although an adaptation effect may partially explain the differences between the CI-pre and control scores the differences may reflect genuine differences, e g , background characteristics, between the two groups A moderate number of cases participated in this study and data were obtained via retrospective measurements Nevertheless, the differences observed between the CI-pre and CI-post situation are fairly substantial Therefore, these limitations do not preclude us from drawing valid conclusions However, further research is necessary to evaluate the validity and reliability of the NCIQ questionnaire

SF-36 scores of the CI study group were compared to the norm scores (n = 2,474) from a Canadian study [17] Canadian scores were very similar to the norm scores (n = 1,063) of a Dutch population sample [16] Table 5 shows that the CI-post scores were very similar to the norm scores on all eight domains Compared to the CI group and the general population-based norm scores, postlingually deaf adults without a CI had substantially poorer scores

on the domains social functioning, role functioning (physical and emotional) and mental health. Especially for policy makers, it may be more informative to compare the effect of CI with other medical interventions. Therefore, results of two transplant studies using the SF-36 are also summarized in Table 5. However, when these instruments are used in different types of disease or medical interventions one should be cautious about the influences of age on the general health status and the influence of co-morbidity conditions. Khan et al.[28] compared three groups of patients using the SF-36: patients with renal failure undergoing haemodialysis, patients receiving peritoneal dialysis and patients who had undergone a renal replacement therapy (transplant). The perception of health in the haemodialysis and peritoneal dialysis patients was significantly poorer than that in the transplant patients and healthy controls. Part of the effect, however, was explained by differences in age and co-morbidity. Rector et al.[29] used the SF-36 to evaluate the impact of a heart transplant on the perceived quality of life. A considerable difference was found between the transplanted patients and the patients on the waiting list for a heart transplant. The average age of the patients on the waiting list and the transplanted patients was 51 years and 53 years, respectively.

Table 5 clearly indicates that a CI has a considerable impact on the perceived HRQoL. On some of the domains the impact was comparable to that of renal transplantation and heart transplantation. In terms of social functioning, for example, the health perception of profoundly deaf patients can be compared to those of patients receiving haemodialysis or patients awaiting a heart transplant. The greatest improvements due to a CI were found for the domains social functioning and role functioning. Not surprisingly, renal or heart transplantation mainly affects physical parameters, but these interventions also have a considerable effect on social functioning, comparable with that of a CI. Apparently, deafness has a greater association with emotional problems than renal or cardiac pathology: a CI had a strong positive effect on emotional problems.

Several studies have made use of the HUI-2 to evaluate quality of life and calculate utilities for different types of condition. Neumann et al.[26] used the instrument on patients suffering from different stages of Alzheimer's Disease (moderate 0.53, mild 0.69). Not unexpectedly, the average age of these patients was relatively high (63 years). Bartman et al.[27] found a HRQoL utility of 0.70 in older patients with intermittent claudication. The mean utility of a group of adult survivors of brain tumours was 0.78. Due to the heterogeneity in this group, utilities ranged from 0.2 to 1. Wyatt et al.[10] found utilities of 0.59 and 0.79 in profoundly deaf adults and in patients fitted with a cochlear implant, respectively. These results are in agreement with our own findings.

The main disadvantage of generic quality of life instruments is their relative insensitivity to some specific health-related aspects of illness. Rector et al.[29] for example found that the SF-36 missed items for health-related distress, sexual dysfunction, problems with sleep and self-image. Dougherty et al.[30] also found that the SF-36 was relatively insensitive to some

clinically important changes in cardiac status and recommended the use of a disease-specific measure such as the Seattle Angina Questionnaire. Similarly, the SF-36 and especially the HUI-2 are insensitive to small changes in the hearing abilities of CI patients. Thus they are unable to evaluate the effect of different types of cochlear implant or speech-coding strategy, even in the same group of patients. Therefore we stress the importance of using a comprehensive disease-specific quality of life instrument, such as the NCIQ in combination with generic instruments.

POLICY IMPLICATIONS

The improved health-related quality of life due to a CI is substantial and will last from the time of implantation until death. Therefore, we conclude that the benefits of a CI expressed in terms of the number of quality-adjusted life years gained may be even greater than those associated with numerous other medical treatments for (non-lethal) diseases.

REFERENCES

1 Blamey P, Arndt P, Bergeron F, et al Factors affecting auditory performance of postlingually deaf adults using cochlear implants Audio Neurootol 1996,1 293-306

2 Cohen NL, Waltzman SB, Fisher SG A prospective, randomized study of cochlear implants New Engl J Med 1993,328 233-237

3 Wexler M, Berliner K, Miller L Psychological effects of cochlear implants patients and index relative perceptions Ann Otol Laryngol 1982,91 59-61

4 Crary W, Berliner K, Wexler M Psychometric studies and interviews with cochlear implant patients Ann Otol Rhinol Laryngol 1982,91 55-58

5 East CA, Cooper HR Extra-cochlear implants the patients viewpoint British Journal of Audiol 1986,20 55-59

6 Tyler RS, Kelsay D Advantages and disadvantages reported by some of the better cochlear implant patients Am J Otol 1990,11 282-289

7 Zhao F, Stephens SDG, Sim SW, et al The use of qualitative questionnaires in patients having and being considered for cochlear implants Clin Otolaryngol 1997,22 254-259

8 Maillet CJ, Tyler RS, Jordan HN Change in the quality of life of adult cochlear implant patients Ann Otol Rhinol Laryngol 1995,165 31-48

9 Wyatt JR, Niparko JK, Rothman ML, et al Cost effectiveness of the multichannel cochlear implant Am J Otol 1995,16 52-62

10 Wyatt JR, Niparko JK, Rothman ML Cost utility of the multichannel cochlear implant in 258 profoundly deaf individuals Laryngoscope 1996,106 816-821

11 Harris JP, Anderson JP, Novak R An outcomes study of cochlear implants in deaf patients audiologic, economic, and quality-of-life changes Arch Otolaryngol 1995,121 398-404

12 Hinderink JB, Krabbe PFM, Broek van den P Development and application of a health-related quality of life instrument for adults with cochlear implants the Nijmegen Cochlear Implant Questionnaire Otolaryng Head Neck (in press)

13 Ware JE, Sherbourne CD The MOS 36-item Short-From Health Survey (SF-36) I conceptual framework and item selection Med Care 1992,30 473-481

14 Aaronson NK, Acquado C, Alonso J, et al International quality of life assessment (IQOLA) project Qual Life Res 1992,1 349-351

15 Brazier JE, Harper R, Jones NMB Validating the SF-36 health survey questionnaire new outcome measure for primary health BMJ 1992,305 160-164

16 Zee van der K, Sanderman R, Heyink J Psychometric properties of the MOS 36-item Short Form Health Survey (SF-36) in a Dutch population (in Dutch) TSG 1993,71 183-191

17 Ware, J E (1996) The SF-36 Health Survey In B Spilker (Ed), Quality of Life and Pharmacoeconomics in Clincial Trials (pp 337-345) Philadelphia Lippincott-Raven Publishers

18 Essink-Bot ML, Krabbe PFM, Bonsel GJ, et al An empirical comparison of four generic health status measures the Nottingham Health Profile, the MOS 36-Item Short-Form Health Survey, the COOP/WONCA charts, and the EuroQol instrument Med Care 1997,35 522-537

19 Torrance GW, Feeny D, Furlong WJ, et al Multiattribute utility function for a comphrensive health status classification system Health Utility Index Mark 2 Med Care 1996,34 702-722

20 Broek van den P, Admiraal R Selection of patients for a cochlear implant Acta Oto-Rhino-

Laryngologica Belg 1991;45: 243-246.
21. Cohen J. Statistical Power Analysis for the Behavioral Sciences. New York: Academic Press; 1977
22. Froberg DG, Kane RL. Methodology for measuring health-state preferences - III: population and context effects. J Clin Epidemiol 1989;42: 585-592.
23. Dolan P. The effect of experience of illness on health state valuations. J Clin Epidemiol 1996;49: 551-564.
24. Breetvelt IS, Dam van FSAM. Underreporting of cancer patients: the case of response-shift. Soc Sci & Med 1991;32: 981-987.
25. Allison PJ, Locker D, Feine JS. Quality of life: a dynamic construct. Soc Sci & Med 1997;45: 221-230.
26. Neumann PJ, Kuntz KM, Leon J, et al. Health utilities in Alzheimer's disease. Med Care 1999;37: 27-32.
27. Bartman BA, Rosen MJ, Bradham DD, et al. Relationship between health status and utility measures in older claudicants. Qual Life Res 1998;7: 67-73.
28. Khan IH, Garratt AM, Cody DJ, et al. Patients' perception of health on renal replacement therapy: evaluation using a new instrument. Neprol Dial Transplant 1995;10: 684-689.
29. Rector TSOSM, Kubo SH. Health status of heart transplant recipients versus patients awaiting heart transplantation: a preliminary evaluation of the SF-36 questionnaire. J Heart Lung Transpl 1993;12: 983-986.
30. Dougherty C.M., Dewhurst T, Nichol WP, et al. Comparison of three quality of life instruments in stable angina pectoris: Seattle Angina Questionnaire, Short Form Health Survey (SF-36), and Quality of Life Index-Cardiac Version III. J Clin Epidemiol 1998;51: 569-575.

CONCLUDING REMARKS

Cochlear implantation has proved itself an effective treatment for total deafness. Thirty years ago, scientists considered successful implantation of the cochlea with long-term stimulation of the auditory nerve to be science fiction. In the meantime considerable experience has been gained with the implantation of postlingual deaf adults, and since 1985 with congenital or early onset deaf children as well.

This treatment has been applied in the Netherlands only since 1985, initially in adults and later in children. The high level of otological and audiological care in the Netherlands combined with a long tradition of education and rehabilitation for the deaf provided good conditions for the application of CI in this country. On the scientific side, a great deal of knowledge of sound perception and speech and language pathology was available. Standardized Dutch hearing and speech perception tests for seriously hearing-impaired and deaf with a cochlear implant were developed at an early stage, enabling the effects of this treatment to be adequately evaluated.

Despite favorable results – which were as good as those produced anywhere else – it still took more than 10 years (and 2 medical-developmental studies) before CI could be registered as a regular form of treatment for total deafness in the Netherlands. The process took even longer for children, which led to a number of them missing a chance for adequate rehabilitation from their deafness. In children it is of great importance that CI be started as early as possible to obtain an optimal result.

This delay in the decision-making process and the resulting relatively small number of implantations performed is the reason the Netherlands is lagging behind other countries in practical experience with CI. A second consequence is that knowledge and experience regarding the technique have inevitably become concentrated among a small number of doctors and rehabilitation experts. These policies have also to some extent damaged scientific development in the Netherlands.

Of the more than 30,000 implantations worldwide, about 1% have been carried out in the Netherlands. Most of the 200 implantations performed in Nijmegen were carried out in postlingual deaf adults and in congenitally or early onset deaf children. The results from CI in prelingual deaf adults were initially less successful for a number of reasons, but even here CI has shown that it can be indicated for specially selected cases. The results of CI in prelingual deaf adults have always been poor as compared to those in postlingual deaf adults. This is one reason why only relatively few implantations have been performed in this group; another is that many of these people traditionally have alternative communication methods at their disposal among family and the deaf community. This led to little interest in the technique within this group; indeed initially there was considerable resistance to CI. The early fear in this group that CI would be a threat to the "deaf culture" or viewed as confirmation that deaf people would not be accepted in society has gradually lessened. The opposite is true:

the better deaf people are integrated in the community, the better they are accepted. CI thus improves their integration.

Although most prelingually deaf CI users obtain poor speech perception results, they are at least able to perceive sound. This can be of great importance performing a signal and warning function or providing support for lipreading. Acoustic contact with the surroundings is of considerable psychological importance. The subjective impression of sound in prelingual deaf people usually proves to be experienced as very positive, despite the low level of objectively measurable results. Use of advanced multichannel implant systems in prelingual deafness appeared to provide no advantage beyond a simple single-channel implant. By contrast, postlingual deaf patients perform much better at speech recognition tests when a multichannel implant with advanced speech coding strategies is used.

At this time cochlear implantation is almost exclusively used in postlingual deafness and in deaf children. However, CI is appropriate for a small number of specially selected prelingual deaf persons. One example is people with Usher's syndrome, who are not only deaf but also suffer from a visual handicap which can lead to blindness.

In the Netherlands, cochlear implantation is carried out at two centres (Utrecht and Nijmegen), each working together with a deaf institute (Effatha, Instituut voor Doven), which is of great importance for the rehabilitation of implanted children.

Due to technological progress in CI and the consequent considerable improvement in results, the indications for use of the technique are gradually shifting toward people with limited residual hearing (90-95 dBHL) who were previously treated with conventional hearing aids but were not able to reach more than 40-50% scores on word perception tests. Better results can be obtained in this group with CI or with a combination of CI and a conventional hearing aid in the contralateral or even in the same ear.[1] The latter is also being investigated using CI with partial electrode insertion combined with a conventional hearing aid, to prevent damage as much as possible to residual nerve fibers. Future improvements in CI technology[2,3] might even further expand the indication for CI. In these cases one should nevertheless remain extremely cautious in view of the risk of losing all residual hearing.

The application of so-called "soft-surgery" techniques, which have been propagated for several years, is said to cause less damage to the still-functioning elements in the inner ear (including the vestibular system) and as a result to further reduce the risk of complications such as further hearing loss or post-operative dizziness.

There have been important developments in the field of diagnostics. In the past few years pre-operative imaging has shown spectacular improvements. It is already possible to distinguish the three separate scalae of the cochlea using special MRI sequences. The auditory nerve can now be imaged. Even more accurate imaging or functional imaging of the neural system, for example with PET scanning (the imaging of metabolic activity)[4] can provide more information about the condition of the auditory nerve and the central auditory system, enabling the optimal determination of electrode placement and thus more predictable results.

Until recently the "direct" effect of CI was measured using tests to measure the perception of sound and speech production. Although the results of these tests are convincing and the advantages of better hearing and a less stigmatising "deaf speech" are obvious, there has recently been a shift of emphasis toward the effects of medical technologies like CI on the subjective experience of the patient. These so-called "quality of life" measurements provide further insight into the "indirect" effects of medical treatment and also make possible a comparison between different treatments for various handicaps or diseases. This gives policy makers, who have to share out the scarce means available in the health service, an instrument to justify their choices to society.

The development of cochlear and other implantation technologies has accelerated in recent years. The development of new electrode arrays that enable placing the electrodes closer and more accurately by the nerve endings ("modiolar hugging") has made new encoding strategies possible. This technology now makes it worthwhile to discover the optimal stimulation pattern for each separate electrode with regard to the specific needs and capabilities of the nerve population within the vicinity of that electrode. Because modiolar hugging reduces the amount of energy required, smaller implant systems can be made.

Making the CI invisible could provide a big advantage. The large, heavy speech processor of a few decades ago was quickly replaced with one the size of a walkman. Recent years have seen the emergence of, and clear demand for, behind-the-ear processors. Further miniaturization would lead to completely implantable systems, with the speech processor and the microphone placed under the skin. This would provide more freedom of movement to people with implants, for instance by making possible swimming with the implant (a frequent cause of complaint regarding present systems among our group of implanted patients).

At the same time work is being done to improve the signal-to-noise ratio using directionally sensitive microphones. A further improvement in the ability to understand speech in noise can be gained by using the binaural capabilities of the brain with bilateral implants.[5,6]

Research into neural growth factors is needed in order to gain more insight into the possibilities for preventing further degeneration and perhaps even (re)generation of the damaged neurones of the auditory nerve and the spiral ganglion. Experiments in this direction have already been carried out in animal models, that demonstrated clear improvement in the survival of ganglion cells.[7] This approach could eventually lead to the pharmacological treatment of deafness. Regaining and enlarging the plasticity of the central auditive systems normally present in young children could be encouraged by the use of neurotrophins, which can lead to the formation of new synapses. Special training methods are already being developed to stimulate this plasticity in children.

Notwithstanding all of these future perspectives for treating deafness, it is obvious that prevention is better than cure. In cases of genetically determined deafness, genetic counselling has a role to play. It may be possible in future to use gene therapy at an early stage of

development to prevent certain forms of deafness. Closer to home, great benefits could be gained through vaccination against certain forms of meningitis, an important cause of deafness in children.

REFERENCES

1. Blamey PJ, Dooley GJ, James CJ, Parisi ES. Monaural and binaural loudness measures in cochlear implant users with contralateral residual hearing. Ear Hear 2000;21:6-17
2. Spelman FA. The past, present and future of cochlear prostheses. IEEE Eng in Med and Biol 1999;18:27-33
3. Clark GM. Cochlear Implants in the third millennium. Am J Otol 1999;20:4-8
4. Hoffman E, Preibisch C, Knaus C, Muller J, Kremser C, Teissl C. Noninvasive direct stimulation of the cochlear nerve for functional MR imaging of the auditory cortex. Am J Neuroradiol 1999;20:1970-1972
5. Green JD Jr, Mills DM, Bell BA, Luxford WM, Tonokawa LL. Binaural cochlear implants. 1992;54:285-294
6. Lawson DT, Wilson BS, Zerbi M, van den Honert C, Finley CC, Farmer JC Jr, McElveen JT Jr, Roush PA. Bilateral cochlear implants controlled by a single speech processor. Am J Otol 1998;19:758-761
7. Marzella PL, Clark GM. Growth factors, auditory neurones and cochlear implants: a review. Acta Otolaryngol 1999;119:407-412

SUMMARY

Chapter 1 provides an introduction to cochlear implant history and technology. Cochlear implantation has developed into a regular and safe treatment for total deafness in adults and children. Only recently has CI been officially recognised in the Netherlands. Protocols have been laid down for the selection, implantation and rehabilitation of the totally deaf in two developmental health-care projects for adults and one project for children. Because the results are continuously improving with advancing technological developments, people with some degree of residual hearing are now also being considered for CI. Development of cochlear implantation has not come to a standstill. Ambitious targets for the rehabilitation of the deaf in the third millennium have been set.

The aim of the present study is to describe results of cochlear implantation in adults regarding the prevention of complications by careful pre-operative evaluation, the effect of CI on prelingually deafened individuals and the results of CI in terms of quality of life.
Chapter 2 describes results and compare advantages and disadvantages of different imaging techniques for the preoperative radiological evaluation of cochlear implant candidates.

Radiological and surgical findings were compared in 100 consecutively implanted patients. All patients underwent high resolution CT scanning of both petrous bones; 28 also underwent MRI. Meningitis was the main cause of deafness (45/100) and the only etiology that caused clinically significant compromised cochlear patency (17/45). CT achieved a Sensitivity of 78% for detecting diffuse cochlear obstruction while MRI achieved 100%. Other abnormalities were detected equally well by CT and MRI imaging, although CT imaging provided more detailed information about bony structures. On the other hand, MRI provided additional information about the internal auditory canal and other possible retrocochlear abnormalities.

We conclude that in cochlear implant candidates with an etiology of meningitis, consideration should be given to starting with MRI rather than CT scanning. The provision of more elaborate information about cochlear patency by MRI facilitates decision-making regarding whether to implant or not and, if so, what implant system to use. In patients with other causes of deafness, a CT scan is sufficient.

Chapter 3: Sixty patients were selected for cochlear implantation and 50 of them received an intracochlear implant (Nucleus). Vestibular function was evaluated before and after surgery using a caloric test and a velocity step test. Sixteen patients had normal or residual vestibular function before surgery, 11 bilateral and 5 unilateral; in 3 of the latter patients, the ear with vestibular areflexia was elected for implantation, which reduced the number of patients at risk for vestibular dysfunction to 13. Vestibular function was preserved in all of these patients except for 4; the risk of vestibular function loss can therefore be rated at about 31%.

Chapter 4 presents individual results of four patients with Usher syndrome type I who

received a cochlear implant. Both single-channel and multichannel implants were used. Because of implant failure, one of the single-channel systems was replaced by a Nucleus multichannel system. Results are compared to the results of five other prelingually deaf cochlear implant users. The performance of the patients with Usher's syndrome on suprasegmental and segmental speech perception tests and on a connected discourse tracking task did not differ significantly from the performance of the other prelingually deaf patients. A significant improvement over time was found at the suprasegmental level for the combined group of Usher's and other patients. No obvious differences were found between the scores from the patients with a single-channel and the patients with a multichannel device. The rehabilitation of the Usher's patients required very little extra effort in comparison with that of the other prelingually deaf patients; all patients reported considerable advantages in hearing abilities and social life.

Chapter 5 presents the evaluation during 2 years follow-up of the auditory and aided lip-reading performance of 8 prelingually and 11 postlingually deaf patients who had received a single or multichannel cochlear implant. The auditory performance was investigated using closed-set pattern and speech recognition tests and a Continuous Discourse Tracking (CDT) task. Although all the patients improved on the pattern recognition level, the most significant improvement was observed in the group of postlinguals who were using a multichannel implant. Only small differences were found between the prelinguals who were using a single or multichannel system and the postlinguals who were using a single-channel system. Comparable results were found on the speech recognition level but the outstanding performance of the postlinguals who were using a multichannel system was even more pronounced. The results on a Continuous Discourse Tracking task were similar; furthermore, speech recognition in the auditory-only condition was only achieved by the postlinguals who were using a multichannel system. On average, the users' evaluations obtained by means of a questionnaire were positive in all the different user groups. It was concluded that it is feasible to implant highly motivated patients who became deaf prelingually and have learnt to use oral-aural communication. Single-channel systems may be as effective as multichannel systems in this group.

Chapter 6 describes the development of a quantifiable, self-assessment health-related quality of life (HRQoL) instrument for use in cochlear implant users. Three principal domains were distinguished: physical, psychological and social. Forty-five postlingually deaf adult multichannel cochlear implant users and forty-six deaf candidates on the waiting list for a CI (control group) participated in the study.

Retrospective scores for the CI group corresponded very well with the scores for the control group. Current QoL scores were substantially higher for all six subdomains. Internal consistency and test-retest reliability coefficients proved to be satisfactory while the ability to detect clinical changes with the NCIQ proved to be good.

The psychometric characteristics of the NCIQ proved to be reliable, probably valid and

sensitive to clinical changes. The data obtained with the NCIQ reflected that the instrument was able to detect that a CI had significant effects on several HRQoL aspects, including the social and psychological domains.

Chapter 7 describes the assessment of the effect of the use of cochlear implants on the health status of postlingually deaf adults. Participants comprised 45 postlingually deaf adult multichannel CI users and 46 deaf candidates on the waiting list for a CI. The latter group acted as controls to corroborate the validity of retrospective completion of the questionnaires by the CI recipients. Three HRQoL instruments were used: 1) a specially developed CI questionnaire (NCIQ), 2) a generic HRQoL questionnaire (SF-36) and 3) a health-state classification system (HUI-2) suited to estimate single preference scores.

Retrospectively estimated pre-implant scores in the CI user group corresponded very well with the scores in the control group. Post-implant scores in the CI users were substantially higher in all six domains ($p < 0.001$) of the NCIQ than the scores in the controls. Effects due to a CI were also observed with the SF-36 in five out of the seven domains ($p < 0.01$). Statistically significant differences between the two groups ($p = 0.001$) were observed in two of the six domains of the HUI-2.

All three questionnaires detected improvements in HRQoL due to CI use. To make a detailed assessment of the effect of a CI on functional outcomes and well-being a special-purpose HRQoL instrument is far more adequate than a general HRQoL instrument. This study also showed that a CI affects several other health domains besides auditory performance. The effect of CI use on general functioning and well-being proved to be considerable.

SAMENVATTING

Hoofdstuk 1: Geeft een introductie in de geschiedenis en de techniek van Cochleaire Implantie. Cochleaire Implantatie (CI) heeft zich ontwikkeld tot een reguliere en veilige behandeling van totale doofheid bij volwassenen en kinderen. Sinds 1982 wordt deze behandeling ook in Nederland toegepast. Gedurende twee ontwikkelingsgeneeskunde projecten werd een uitgebreid protocol ontwikkeld voor de selectie, implantatie en revalidatie van totaal dove volwassenen.

Onder andere dankzij technologische ontwikkelingen zijn de resultaten van de behandeling in de loop der tijd steeds verbeterd. Ook mensen met een zekere mate van restgehoor komen tegenwoordig in aanmerking voor CI. De ontwikkelingen op het gebied van Cochleaire Implantatie gaan nog steeds door. Voor het derde millennium zijn ambitieuze doelen geformuleerd.

Het doel van de huidige studie is een beschrijving te geven van de resultaten van Cochleaire Implantatie bij volwassenen met betrekking tot het voorkomen van complicaties door zorgvuldige preoperatieve evaluatie, het effect van CI bij prelinguaal dove volwassenen en de resultaten van CI in het kader van Kwaliteit-van-Leven.

In hoofdstuk 2 worden de resultaten besproken van verschillende radiologische beeldvormende technieken voor de preoperative evaluatie van CI kandidaten. De radiologische en chirurgische bevindingen van 100 opeenvolgend behandelde CI patienten werden vergeleken. Alle patienten ondergingen een CT onderzoek van beide oren; bij 28 werd ook een MRI onderzoek verricht. De belangrijkste oorzaak van doofheid was meningitis (45/100) en tevens de enige etiologie waarbij klinisch relevante obstructie van de cochlea werd gevonden (17/45). De sensitiviteit van CT voor het opsporen van diffuse cochleaire obstructive was 78% terwijl die voor MRI 100% bedroeg. Andere afwijkingen werden even goed gezien op CT en MRI scans hoewel op CT scans een meer gedetailleerd beeld van benige structuren werd verkregen. Aan de andere kant gaven de MRI scans extra informatie over de inwendige gehoorgang en andere mogelijke retrocochleaire afwijkingen. Concluderend lijkt het zinvol om bij CI kandidaten die doof werden door meningitis te beginnen met een MRI scan in plaats van met een CT scan. De extra nauwkeurige informatie die een MRI scan geeft met betrekking tot de doorgankelijkheid van de cochlea maakt het makkelijker in een vroeg stadium te besluiten om al dan niet door te gaan met de selectieprocedure voor CI en zo ja voor welk implant systeem dan het beste kan worden gekozen. Bij patienten met een andere doofheids oorzaak kan volstaan worden met alleen een CT scan.

In hoofdstuk 3 wordt de vestibulaire functie geevalueerd bij 50 patienten die een intracochleaire Nucleus implant geplaatst kregen. Pre - en postoperatief werd een calorisatie test en een draaistoel onderzoek verricht. Bij 16 patienten werd preoperatief een normaal of gedeeltelijk functionerend vestibular systeem vastgesteld, bij 11 bilateraal en bij 5 unilateraal; bij 3 van deze laatste patienten werd het oor met areflexie uitgekozen voor implantatie zodat

13 patienten overbleven die een mogelijk risico liepen op vestibulaire beschadiging door implantatie. De vestibulaire functie bleef in 9 van deze patienten behouden. Het risico op uitval van de vestibulaire functie door CI zal dan ook ongeveer rond de 31% liggen.

In hoofdstuk 4 worden de resultaten gepresenteerd van vier patienten met het syndroom van Usher type I met een cochleaire implant. Zowel een- als meerkanaals impants waren gebruikt. Vanwege een technische storing was bij een van deze patienten een eenkanaals implant vervangen door een Nucleus meerkanaals implant. De resultaten zijn vergeleken met die van vijf andere prelinguaal dove CI gebruikers. De resultaten die de Usher patienten behaalden op suprasegmentele en segmentele spraakperceptie tests en op een spraakafzienstest (CDT test) verschilden niet significant van de resultaten die door de andere prelinguaal dove patienten werden behaald. Er werd een significante verbetering van de resultaten gezien over de tijd voor wat bereft de resultaten op de suprasegmentele tests voor de gehele groep onderzochte patienten. Er werden geen duidelijke verschillen waargenomen tussen de patienten met een- en de patienten met een meerkanaalssysteem. De revalidatie van de patienten met het syndroom van Usher vereiste weinig extra inspanning in vergelijking met de andere prelinguaal doven; Alle patienten rapporteerden aanzienlijke voordelen van CI met betrekking tot de geluidswaarneming en de sociale omgang.

In hoofdstuk 5 worden de geluids- en spraakperceptie resultaten met en zonder spraakafzien geevalueerd van 8 prelinguaal en 11 postlinguaal doven die met een een- of meerkanaals implant werden behaald gedurende 2 jaar na implantatie. De mate van geluidsperceptie werd onderzocht middels gesloten patroon- en spraakherkennings tests en middels een spraakafziens test (CDT). Hoewel bij alle patienten een verbetering werd gezien op het patroonherkennings niveau, werd de meeste vooruitgang gezien bij de postlinguaal doven met een meerkanaalsimplant. Tussen de resultaten van de prelinguaal doven met een een- of meerkanaals implant en de postlinguaal doven met een eenkanaals implant werden slechts geringe verschillen waargenomen.

Vergelijkbare resultaten werden gevonden voor het spraakperceptie niveau maar hier vielen de goede resultaten die door de postlinguaal doven met een meerkanaals implant werden behaald nog duidelijker op. De resultaten op de CDT test lieten eenzelfde beeld zien. Open spraakverstaan werd slechts gezien in de groep postlinguaal doven met een meerkanaals system. De subjective "gebruikersbevindingen" die gemeten werden middels een vragenlijst waren over het algemeen positief voor alle onderzochte groepen. Als conclusive kon worden gesteld dat CI haalbaar is bij goed gemotiveerde prelinguaal dove volwassenen met een orale communicatie achtergrond. Eenkanaals cochleaire implants zijn in deze groep waarschijnlijk even effectief als meerkanaals implants.

In hoofdstuk 6 wordt de ontwikkeling en eerste toepassing beschreven van een kwantificeerbaar "self-assessment" gezondheids gerelateerd Kwaliteit-van-Leven instrument voor gebruik bij cochleaire implant (CI) gebruikers.

Er werden drie hoofd domeinen onderscheiden: fysiek, psychisch en sociaal. 45

postlinguaal dove volwassenen met een meerkanaals implant en 46 poslinguaal dove volwassenen van de CI wachtlijst (controle groep) deden mee met de studie.

De retrospective scores van de CI groep kwamen goed overeen met die van de controlegroep. De huidige Kwaliteit-van-Leven scores waren beduidend hoger voor alle zes de subdomeinen. De interne consistentie en de test-hertest betrouwbaarheid bleken voldoende terwijl de gevoeligheid van de vragenlijst om klinische veranderingen aan te tonen goed bleek te zijn. Geconcludeerd werd dat de psychometrische eigenschappen van de NCIQ betrouwbaar, waarschijnlijk valide en gevoelig voor klinische veranderingen zijn. De met de NCIQ verkregen gegevens toonden aan dat CI een belangrijke invloed heeft op verschillende gezondheids gerelateerde Kwaliteit-van-Leven aspecten waaronder sociale en psychologische.

In hoofstuk 7 wordt het effect van CI op de gezondheidstoestand van postlinguaal dove volwassenen geevalueerd. 45 postlinguaal dove volwassenen met een meerkanaals implant en 46 postlinguaal dove volwassenen van de wachtlijst voor CI werden geincludeerd in de studie. De laatste groep diende als controlegroep om de validiteit van de retrospective gegevensinzameling te versterken. Er werden 3 gezondheids gerelateerde Kwaliteit-van-Leven instrumenten gebruikt: 1) een specifiek voor CI ontwikkelde vragenlijst (NCIQ), 2) een generieke Kwaliteit-van-Leven vragenlijst (SF-36) en 3) een gezondheidstoestand klassificatie system (HUI-2) geschikt om de gezondheidstoestand in een enkel getal weer te geven. De retrospectief aangegeven preimplantatie scores in de CI groep kwamen zeer goed overeen met de scores in de controlegroep. De postimplant scores op de NCIQ waren significant hoger ($p< 0,001$) dan de scores in de controlegroep. Effecten door CI werden ook waargenomen met de SF-36 in 5 van de 7 domeinen ($p< 0,01$). Statistisch significante verschillen tussen de 2 groepen ($p< 0,001$) werden ook gezien in 2 van de 6 domeinen van de HUI-2. Geconcludeerd werd dat alle drie de vragenlijsten verbeteringen door CI konden meten. Voor een gedetailleerde evaluatie van de effecten van CI op het functioneren en het algeheel welbevinden is een ziekte-specifiek Kwaliteit-van Leven instrument veel geschikter dan een meer generiek Kwaliteit-van-Leven instrument. Deze studie liet tevens zien dat CI van invloed is op veel meer gezondheidsdomeinen dan alleen het auditief functioneren. Het effect van CI op het algeheel functioneren en het welbevinden bleek bovendien aanzienlijk.

DANKWOORD

In het kader van meerdere ontwikkelingsgeneeskunde projecten is uitgebreid onderzoek gedaan naar de effecten van Cochleaire Implantatie. Aan dit onderzoek hebben veel mensen hun bijdrage geleverd. Ik ben dankbaar voor het feit dat ik in staat ben gesteld tijdelijk deel te mogen uitmaken van dit Cochleaire Implant team en in deze functie aan dit promotieonderzoek heb mogen werken. De eersten die ik wil bedanken zijn echter mijn ouders en mijn gezin die mij altijd hebben gesteund en gemotiveerd tijdens studie en promotie en mijn bij tijd en wijle wat mindere humeur na een zoveelste correctie steeds hebben vergeven. Eenzelfde dankbaarheid voel ik jegens Paul van den Broek, mijn promotor die mij meestal met zachte hand en eindeloos veel geduld en zelden met minder zachte hand heeft gemotiveerd om de vaart erin te houden (krijgen). Daarnaast heeft hij altijd een belangrijke kritisch inhoudelijke bijdrage aan het onderzoek en de publicaties geleverd. Mijn co-promotor Lucas Mens vormde een belangrijke steun en toeverlaat bij het tot stand komen van meerdere publicaties. Op hem kon ik altijd rekenen voor uitleg en voor een steeds frisse en creatieve kritiek op mijn werkstukken. Teun Spies, mijn voorganger op het CI project wil ik bedanken voor de verhelderende inwerkperiode en zijn nalatenschap van vele keurig gerubriceerde en ingebonden literatuurstudies. Andere mensen die nauw betrokken waren bij de Cochleaire Implant projecten en zonder wie ik dit boekje nooit had kunnen schrijven zijn Andy Beynon, die verreweg de meeste gehooronderzoeken bij de CI kandidaten en gebruikers heeft afgenomen, Ad Snik, die mij vaak met raad en daad in het onderzoek heeft bijgestaan en Sonja van Oosterhout, die een belangrijke bijdrage heeft geleverd als secretaresse van het project en Frank Joosten, die tijd noch moeite heeft gespaard om alle radiologische gegevens met mij door te nemen. Hetzelfde geldt voor de medewerkers van het Cochleaire Implant team van het Instituut voor Doven waarvan ik met name José van Grinsven-Verbakel, Margreet Peters-Bos, Jacqueline Velthoven-van der Harten, Jan Brokx en Jef Claassen wil noemen.

Een speciaal woord van dank gaat uit naar Ruben Alvarado, die het belangrijkste deel van het edit- en layoutwerk alsmede enig vertaalwerk heeft verricht en hiervoor namens mij de strijd met de soms ondoorgrondelijke eigenzinnigheid van de verschillende tekstverwerkers is aangegaan en in mijn voordeel heeft beslist.

Last but not least een dankbetuiging aan alle CI-kandidaten en gebruikers die zonder al te veel te mopperen steeds hebben meegewerkt aan de vele onderzoeken, tests en vragenlijsten die op hen zijn losgelaten. Allen dank.

CURRICULUM VITAE

De auteur is geboren op 10 april 1962 te Den Haag. In 1982 behaalde hij het Atheneum diploma aan het Groen van Prinsterer College te Den Haag. In datzelfde jaar werd begonnen met de studie geneeskunde te Leiden. 1987 en 1988 werden gewijd aan de wetenschap in de vorm van een wat uit de hand gelopen doctoraalstage waarbij onderzoek werd gedaan naar de vroege ontwikkeling van het centraal zenuwstelsel in de rat, die gevolgd werd bij de Leidse Neuroregulatie groep onder leiding van (inmiddels Prof.) Dr. E. Marani en (inmiddels Dr.) E.A.J.F. Lakke. Na de hierop volgende co-schappen werd meer klinische ervaring opgedaan als AGNIO chirurgie in het Dordrechtse Merwede Ziekenhuis. In 1992 begon hij bij de afdeling KNO van het Academisch Ziekenhuis Nijmegen als coördinator van het tweede ontwikkelingsgeneeskunde project cochleaire implantatie bij volwassenen. Van 1993 tot 1998 volgde hij hier ook de A-opleiding tot KNO-arts bij Prof. Dr. P. van den Broek. De B-opleiding werd in het Rijnstate Ziekenhuis in Arnhem gevolgd bij Dr. E.R. Soudijn. Sinds 1998 is hij werkzaam in de KNO-maatschap Winterswijk in associatie met J.R. Alvarado-van Os in het Streekziekenhuis Koningin Beatrix.

Publication of this thesis was financially supported by:

AstraZeneca BV, Stichting Atze Spoor Fonds, Entermed BV, Claxo Wellcome BV, L. de Haan Hoorapparaten, Horen Audiciens, Mediprof Medical Products, Oticon Nederland BV, GN Resound BV, Schering-Plough BV, Smith&Nephew Nederland BV, Veenhuis Medical Audio BV.

Stellingen

Behorende bij het proefschrift
Cochlear Implants In Adults:
Results From The Nijmegen-Sint Michielsgestel
Cochlear Implant Program

Nijmegen 26 maart 2001
JB Hinderink

1. Bij een CI kandidaat die doof werd door meningitis verdient het aanbeveling een MRI scan van de ossa petrosa te laten maken om optimaal geïnformeerd te zijn over de doorgankelijkheid van de cochlea. *(dit proefschrift)*

2. De uitkomst van het preoperatief verrichte evenwichtsonderzoek is soms van doorslaggevende betekenis voor het maken van de keuze voor het te implanteren oor. *(dit proefschrift)*

3. Cochleaire Implantatie kan in bijzondere gevallen bij prelinguaal dove volwassenen worden toegepast. Dit geldt in het bijzonder voor prelinguaal doven met een additionele visuele handicap zoals bij het syndroom van Usher. *(dit proefschrift)*

4. Een ziekte-specifiek kwaliteit van leven instrument zoals de Nijmegen Cochlear Implant Questionnaire biedt een waardevolle aanvulling op de bestaande psychometrische tests. *(dit proefschrift)*

5. De gevolgen van éénorigheid bij kinderen worden vaak onderschat. Indien de (functionele) éénorigheid het gevolg is van een gehoorgangatresie of van chronische middenoor problemen dient overwogen te worden een BAHA hoortoestel toe te passen, eventueel tijdelijk.

6. Bij de keuze van een multi-slice multi detector CT scanner speelt naast snelheid en beeldkwaliteit, stralenbelasting en -beperking een belangrijke rol.

7. De ordening van ten minste de topologische projecties in het centraal zenuwstelsel kan worden verklaard uit de spatiële interactie van rijpingsgradiënten in de bron en doel gebieden. *(Leergaard TB, Lakke EAJF, Bjaalie JG. J Comp Neurol, 1995)*

8. Het is typerend voor de huidige gezondheidszorg dat men voortdurend spreekt van werklast in plaats van werklust.

9. Kwaliteit van zorg staat in geen verhouding tot kwaliteit van leven

10. Niet alle supratip depressies zijn psychogeen

11. Een oude achterhoekse volkswijsheid geeft blijk van een gezonde kijk op het omgaan met ziekten en handicaps: " 't is vanzelf gekaome; 't zal ook vanzelf weer overgaone". De wijsheid zit hem in de zin die hierna meestal volgt: "en anders maor nie".